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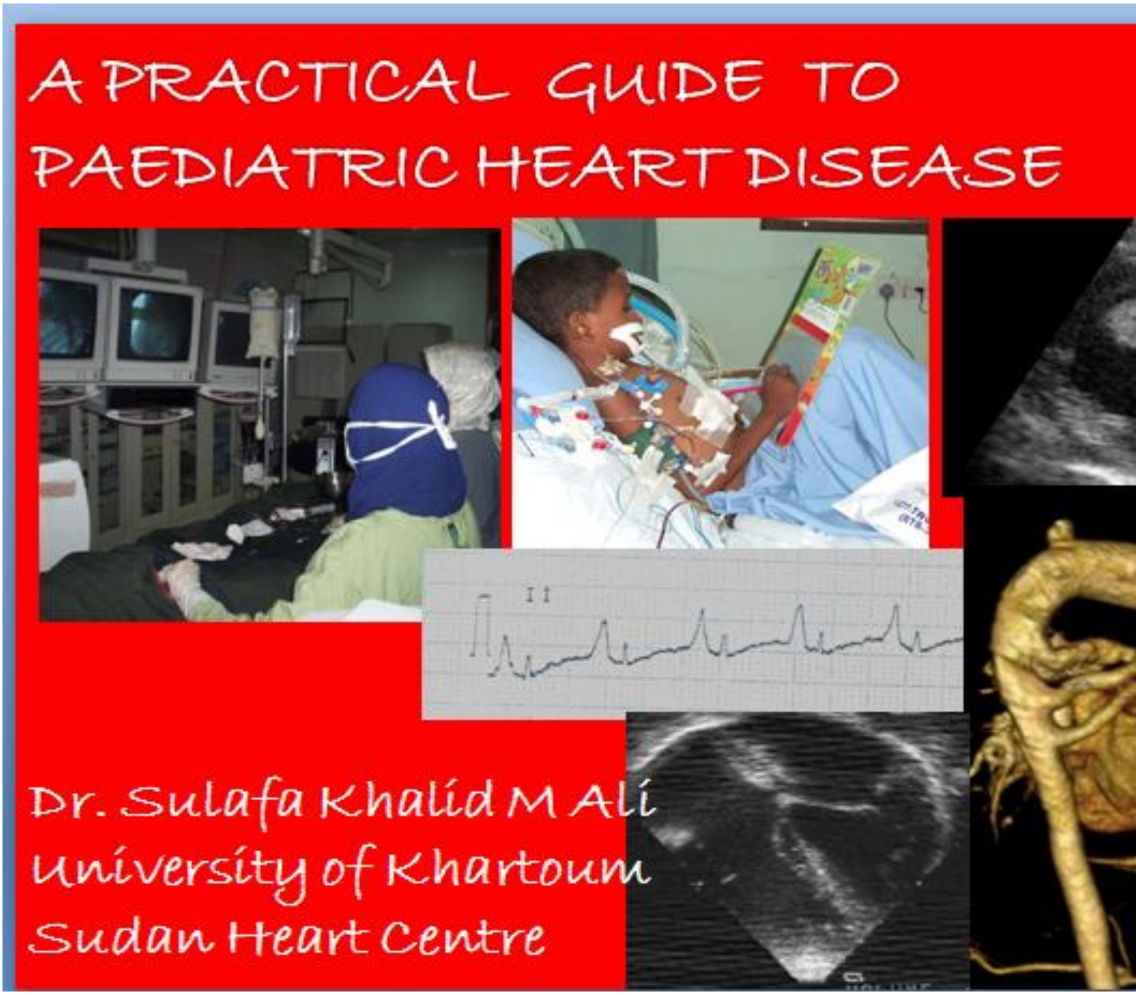

A PRACTICAL GUIDE TO PAEDIATRIC HEART DISEASE

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Contents

Chapter 1
Clinical Evaluation of Children with Heart Disease

1.1 History taking 8
1.2 Physical Examination 11

Chapter 2
Investigations of Children with Heart Disease

2.1 Chest X Ray and other Radiological Investigations 22
2.2 Paediatric Electrocardiogram Interpretation 31
2.3 Basic Echocardiography 38
2.4 Cardiac Catheterization 44

Chapter 3:
Approach to Common Presentations of Paediatric Heart Disease: Case Scenarios

General Approach 50
Scenario 1: Infants with Congestive Heart Failure and Normal Oxygen Saturation 53
Scenario 2: Low Oxygen Saturation without Heart Failure 60
Scenario 3: Low Oxygen saturation and Heart Failure 67
Scenario 4: The sick Neonate 77
Scenario 5: A Sound in My Child's Heart (murmurs) 84
Scenario 6: Eisenmenger's Syndrome 87
Scenario 7: Acute Rheumatic Fever /Rheumatic Heart Disease 91
Scenario 8: Myocarditis and Cardiomyopathy 99
Scenario 9: Infective Endocarditis 109
Scenario 10: Kawasaki Disease 112

Scenario 11: Pericarditis 115
Scenario 12: Supraventricular Tachycardia 118
Scenario 13: A Baby with Bradycardia 122
Scenario 14: A Child with Syncopal Attacks 125
Scenario 15: Involvement of the Heart in Connective Tissue Diseases ... 130
Scenario 16: Congenital Heart Disease in Adults 133
Scenario 17: The Metabolic Syndrome 138

Chapter 4
The difficult made easy

4.1 Complex Congenital Heart disease ....
4.2 Congenital Heart Surgery ....
4.3 The Heterotaxy Syndrome ....
4.4 Ethical Issuues in Paediatric Cardiology ....

Appendices

Commonly Prescribed Medications ....
Paediatric Basic and Advanced Life Support ....
Normal Echo Values ....
Common Syndromes Associated with Heart Diseases ....
Body Mass Index Charts ....
Waist Circumference Reference Values ....

Preface:

The last decades witnessed remarkable developments in the field of Paediatric Cardiology. Diagnostic methods were revolutionized due to improved ultrasound and magnetic resonance technology; consequently cardiac catheterization underwent a major turn from diagnostic to interventional roles. Refinements of cardiac surgical techniques for neonates, infants and children with simple and complex anatomy changed the outlook for many patients with congenital heart disease.

To cope with these advancements medical students as well as practicing physicians need a practical reference for the basics of paediatric cardiology.

The first part of this book details the clinical evaluation of infants and children with heart disease, the second part gives a brief review of cardiac investigations and the third chapter addresses common cardiac problems. In the last section we discussed topics that are generally found difficult by physicians. Most of the figures belong to our patients. As much as we could, an evidence based approach was adopted with citation of updated references from the rapidly growing literature. We emphasized the role of clinical judgement and the rationale utilization of available technology.

I would like to express my gratitude to my teachers, my colleagues and my students who teach, practice and study in a challenging environment and yet, still able to have an admirable performance and give us a lot of inspiration.

We pray to God that this work will be accepted for the benefit of our colleagues and our children.

Dr.Sulafa Khalid M Ali
Khartoum in April 2010

Dedication

To my family: you gave me your own time that I spent writing this book, your support and understanding which made this work a reality.

To my patients: you were the inspiration!

Abbreviations:

AR: aortic regurgitation
AS: aortic stenosis
ASD: atrial septal defect
AV: aortic valve
BP: blood pressure
Cath: catheterization
CHD: congenital heart disease
CHF: congestive heart failure
COA: coarctation of the aorta
CT: Computerized tomography
CXR: chest Xray
DCM: dilated cardiomyopathy
ECG: electrocardiograph
Echo: echocardiography
ESM: ejection systolic murmur
PSM: pansystolic mrmur
HLHS: hypoplastic left heart syndrome
HOCM: hypertrophic obstructive cardiomyopathy
IE: infective endocarditis
LA: left atrium
LV: left ventricle
LVH: left ventricular hypertrophy
MR: mitral regurgitation
MRI: magnetic resonance imaging
MS: mitral stenosis
MV: mitral valve
O2 sat: oxygen saturation
PA: pulmonary artery
PAPVD: partial anomalous pulmonary venous drainage
PDA: patent ductus arteriosus
PHTN: pulmonary hypertension
PR: pulmonary regurgitation
PS: pulmonary stenosis
RA: right atrium
RAD: right axis deviation
RBBB: right bundle branch block

RV: right ventricle
RVH: right ventricular hypertrophy
S1: first heart sound
S2: second heart sound
SVT: supraventricular tachycardia
TAPVD: total anomalous pulmonary venous drainage
TGA: transposition of the great arteries
TOF: Tetralogy of Fallot
TR: tricuspid regurgitation
VSD: ventricular septal defect
WPW: Wolff-Parkinson-White

Chapter 1

Clinical Evaluation of Children with Heart Disease

1.1 History Taking:

- Communicate with the patient and family: introduce yourself.

Table 1:
Common presenting symptoms and causes of referral:

Neonates	Infants	Older children
Respiratory distress Cyanosis Murmurs Neonatal collapse Abnormal antenatal ultrasound Abnormal CXR Family history of heart disease Dysmorphic syndromes	Recurrent chest infections Respiratory distress Failure to thrive Cyanosis Murmurs Kawasaki disease Systemic disease Abnormal CXR Family history of heart disease Dysmorphic syndromes	Rheumatic fever Murmurs Respiratory symptoms Oedema Palpitations Effort intolerance Syncope Chest pain Systemic disease Abnormal CXR Family history of heart disease Dysmorphic syndromes

- Pregnancy: ask about exposure to teratogenic agents e.g. medications like anticonvulsants, radiation. Maternal disease: fever and skin rashes could indicate viral infections like **to**xoplasmosis , **r**ubella, **c**ytomegalovirus and **h**erpes (TORCH). Polyhyrdamnios can indicate foetal CHF which may result from fetal cardiomyopathy or arrhythmias. Maternal diabetes mellitus is associated with CHD like TGA, but commonly with benign asymmetric septal hypertrophy. Antenatal ultrasound is an important tool of antenatal diagnosis of CHD, ask if it was done and its result. Mothers should be advised about preconception folic acid use which had been proved to decrease the incidence of serious CHD,

- Birth: cyanosis, respiratory distress and murmurs discovered at birth. Emergency caesarean section is often performed for mothers due to fetal bradycardia related to congenital heart block.
- Cyanosis: when mild can be missed by parents. Ask about cyanotic spells characterised by: increasing cyanosis, sudden respiratory distress and limpness (loss of tone). In older children inquire about squatting during exertion. These symptoms suggest TOF and TOF-like conditions.
- Respiratory difficulty: There are two types of respiratory distress:
  A: Chronic respiratory distress:
  Indicates CHF or chronic lung disease (rarer causes include chronic metabolic acidosis).
  B: Episodic respiratory distress:
  When associated with cyanosis indicates 'cyanotic spells'. Respiratory causes of episodic distress include bronchial asthma.
  Respiratory distress is an almost constant feature of CHF in infants. The timing of respiratory distress indicates the type of lesion as CHD have a specific chronological sequence of presentation.
- Feeding: interrupted feeding is an important symptom of CHF in infant, it is equivalent to exercise intolerance experienced by older children and adults with CHF. It is usually associated with frontal sweating which results from sympathetic compensatory activity.

Poor weight gain is a sensitive indicator of CHF in infants; it is due to the following:

1. **Increased energy expenditure secondary to chronic respiratory distress, increased heart rate and recurrent respiratory infections.**
2. **Decreased nutrient intake secondary to interrupted feeding, inability to take large volumes and decreased gastrointestinal absorption.**

- Recurrent respiratory infections: common presentation of CHF due to increased pulmonary blood flow, segmental lung collapse secondary to compression of the bronchial tree by the enlarged pulmonary arteries and heart chambers. In addition patients with CHD are prone to infections because of their growth deficiency and hypotonia.
- Effort intolerance : in infants this is manifested as interrupted feeding while in older children it can be quantified by the **New York Heart Association (NYHA)Classification** :
  NYHA Class I: Asymptomatic
  NYHA Class II: Symptoms on severe exertion
  NYHA Class III: Symptoms on mild exertion
  NYHA Class IV: Symptoms at rest
  Special scores are used to quantify CHF in infants (see page 19)
- Syncope: The most common cause is vasovagal syncope (simple postural hypotension). Cardiac causes include serious arrhythmia (e.g. ventricular tachycardia) and severe left ventricular outflow tract obstruction e.g. AS and HOCM.An important cause of syncopy is pulmonary hypertention (primary or secondary). Detailed history of the event should include the time of the event (during

standing or not), association with exertion or emotional changes and the description of the event. The typical scenario in vasovagal syncope is blackout on standing and immediate relief on lying flat. The following associations should raise concern about serious arrhythmias, (commonly associated with long QT syndrome), cardiomyopathy or other serious heart disease :

  - Family history of sudden cardiac death.
  - Prolonged loss of consciousness especially if associated with pallor, cold extremities, bradycardia or need for resuscitation.

  The differential diagnosis includes epilepsy syndromes.

- Chest pain: in the paediatric age group it is rare to have a cardiac cause for chest pain although it is a common symptom. The most common cause is muscular pain, costochondritis, pleurisy or psycho-somatic causes. Cardiac causes of chest pain include pericardities and rarely myocardial ischemia which can be associated with severe AS and HOCM. Coronary artery disease is rare in the pediatric age group and can be due to Kawasaki disease, congenital coronary artery anomalies and early atherosclerosis in familial hypercholesterolemia.
- Palpitations: common causes of palpitations include anxiety especially in teenagers. Non cardiac causes like anaemia and hyperthyroidism need to be considered. The commonest cardiac cause of palpitations is SVT. Features that help to differentiate SVT from non- cardiac causes of palpitations include:
  - Abrupt onset and cessation.
  - A very fast rate.
  - Association with feeling unwell, dizzy or sweaty.
- Joint pain and swelling: migratory (flitting) arthritis involving large joints indicates acute rheumatic fever

while chronic non-migratory arthritis points to connective tissue diseases which can be associated with cardiac involvement like MR , AR and PHTN . Other manifestations of acute rheumatic fever like skin nodules, skin rash and abnormal movements should be carefully sought.

- Neurological and developmental history: Neurological symptoms include:
    - Hemiparesis that could result from cerebral infarction due to hyperviscocity in patients with cyanotic CHD, thrombo-embolism in cardiomyopathy, endocarditis or following open heart surgery.
    - Convulsions and symptoms of increased intracranial pressure: Can be associated with brain abscess in patients with cyanotic CHD.
    - Abnormal movements: Chorea is involuntary purposeless movements involving the trunk and limbs associated with emotional liability. It is a major criterion of acute rheumatic fever.

  -Many syndromes with delayed development are associated with CHD (see appendix IV), therefore documentation of neuro-developmental state is very important.
- Family history: Genetic cardiac diseases include cardiomyopathies (dilated, hypertrophic and noncompaction), long QT syndrome and other inherited syndromes as well as CHD.
- Social history including the impact of heart disease on the child’s activity and family social and financial issues.

1.2 Clinical Examination of *Infants and Toddlers* with Heart Disease:

The 'G*olden Rules* 'for examining any patient:

1. **Good communication with patient and family**
2. **Minimum disturbance**

Use inspection to get most of the information.
Leave the patient on the mother's lap.
The first sign to look for is: *Respiratory distress*

Look for:

- Respiratory distress: count the respiratory rate and note the presence of active alae nasi, use of accessory muscles of respiration, intercostals and subcostal retractions.
- Colour (pink, pale, blue): the nail bed is the best place to look for peripheral cyanosis which indicates the presence of central cyanosis. Later (after auscultation) attempt to look at the oral cavity for central cyanosis. Cyanosis can easily be missed by the eye therefore pulse oximetry should be done to *all* patients suspected to have cardiac disease. The normal O2 sat is >95%, O2 sat 85- 94% is usually not detected by the eye.
- Clubbing: Usually starts after 6 months of age. The best place to look for clubbing is the big toe. It is an indirect indicator of cyanosis so pulse oximetery is mandatory. If oxygen saturation is normal non cardiac causes are likely.
- Note the growth status in relation to age (normal growth or failure to thrive)

- Dysmorphic features : common associations: Down’s, Noonan’s Di-George, William’s and Turner’s syndromes and other chromosomal disorders, associations like VACTERL (*V*ertebral anomalies, *a*nal atresia,*c*ongenital heart defects, *t*racheo*e*sophageal (TE) fistula,*r*enal dysplasia, *l*imb anomalies) and CHARGE (*C*oloboma, *h*eart defects, choanal *a*tresia, growth or mental *r*etardation, *g*enitourinary anomalies, *e*ar anomalies, genital hypoplasia) associations.(see appendix VI for common syndromes)
- Growth parameters: Get the weight, length and head circumference and plot them on the appropriate centile charts. Remember that in developing countries failure to thrive is commonly related to decrease nutrient intake, this often confuse the picture of infants with moderate/small shunts.

Undress the patient *with the minimum disturbance* and look for:

- Chest deformity: Harrison's sulci: the commonest deformity seen in infants with chronic CHF. There is symmetrical bilateral depressions of the middle part of the lower ribs causing the sternum to look bulged.

- The precordium: prominent pulsations can be seen at the apex indicating LV or epigastric area indicating RV enlargement.
- Surgical scars:
  - Median sternotomy: all open heart and some closed heart procedures like Blalock-Taussig shunt are done through a median sternotomy approach.

- Lateral thoracotomy: closed heart procedures like repair of COA, Blalock-Taussig shunt and PDA ligation are done through this incision.

- Palpate the precordium for:
  1. The apex beat (the most inferior and lateral point at which the cardiac pulsations can be distinctly felt).If it is displaced it indicates LV enlargement.
  2. Thrills and their locations: indicate that the murmur is grade 4 or more.
  3. Parasternal and epigastric heaves: indicate RVH.
  4. Prominent second heart sound in the pulmonary area: indicates pulmonary hypertension.
  5. The peripheral pulses: palpate the brachial pulse for the rate and rhythm. (Table 2 shows the normal heart rate in different age groups). Right and left brachial pulses should be compared, CoA can be associated with diminished left brachial pulses due to involvement of the left subclavian artery.Variation of the pulse rate with breathing (sinus arrhythmia) is a common normal finding (Fig1). The quality of the femoral pulse (normal, weak or absent) should be carefully examined, this can cause a lot of anxiety to young infants and if the physician can palpate the *posterior tibial or dorsalis pedis pulse* COA is effectively ruled out. Radio-femoral delay is not usually looked for in infants because of the rapid heart rate.Pulsus paradoxus (paradoxical pulse) is suspected when there is marked variation in the volume of arterial pulses with the respiratory cycle indicating an exaggerated fall of BP during inspiration. It is associated with pericardial tapenade and severe bronchial asthma.

***Always* check femoral pulses in the neonate, absent or week femoral pulse indicates the possibility of left sided obstructive lesions (commonly COA).**

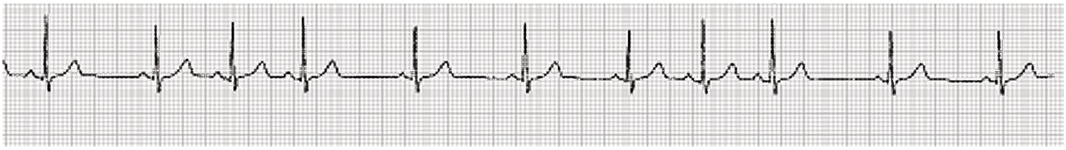


Figure 1
ECG rhythm strip showing respiratory sinus arrhythmia. RR interval slows in expiration and fastens on inspiration with a normal p wave preceding each QRS.

Table 2
Normal heart rate at different age groups

Age	Mean (range)
Newborn	145 (90–180)
6 months	145 (105–185)
1 year	132 (105–170)
4 years	108 (72–135)
14 years	85 (60–120)

6. Palpate for the liver and check the liver span: a common source of erroneous diagnosis of CHF is a liver that is pushed down by a hyperinflated right lung causing false hepatomegaly. The spleen can be enlarged in infants with CHF and in patients with IE.

Causes of Large Volume Pulse

1. **Large PDA**
2. **Hyper dynamic circulation (fever, anaemia).**
3. **Severe AR.**
4. **Truncus arteriosus**
5. **Large aorto-pulmonary window.**
6. **Arterio-venous malformations**

Causes of Small Volume Pulse

1. **Cardiogenic shock: severe cardiomyopathy/myocarditis.**
2. **Hypovolemic and late septic shock.**
3. **Left sided obstructive lesions like AS and HLHS.**

Auscultation:

You may choose to auscultate before palpation to avoid disturbing the infant.

- Auscultate the four basic areas (see Fig 2) plus 3 *special areas*:
  1. The infraclavicular area for PDA
  2. The second aortic area for sub AS and AR.
  3. The back: Murmurs heard at the back are:

i. Continous murmur of aorto pulmonary collaterals in patients with pulmonary atresia.
ii. Ejection systolic murmur of adult-type COA

- Listen to S1: usually normal in children.
- S2: Prominent (PHTN), wide fixed splitting (ASD), and single: TGA and pulmonary atresia.

Murmurs:

(Figs 2 and 50).

- Grading of murmurs:
  Grade 1 Barely audible
  Grade 2 Soft, but easily audible
  Grade 3 Moderately loud, but not accompanied by a thrill
  Grade 4 Louder and associated with a thrill
  Grade 5 Audible with the stethoscope barely on the chest
  Grade 6 Audible with the stethoscope off the chest
- Other sounds (S3,S4, friction rub, clicks)

Murmurs Heard Below the Nipple Line

- **Inflow murmurs (murmurs arising from the two AV valves i.e. the mitral and tricuspid): MR, MS and TR.**
- **VSD.**
- **Commonly *pansystolic*.**

Murmurs Heard Above the Nipple Line

- **Outflow murmurs (arising from aortic and pulmonary valves) and ASD.**
- **Usually *ejection systolic* (AS, PS and ASD)**
- **Early diastolic murmurs (AR, PR)**

Radiation:

- Systolic apical murmurs radiate to the axilla (MR), aortic stenosis murmur to the neck, and pulmonary stenosis (especially pulmonary artery branch stenosis) murmur to the back.

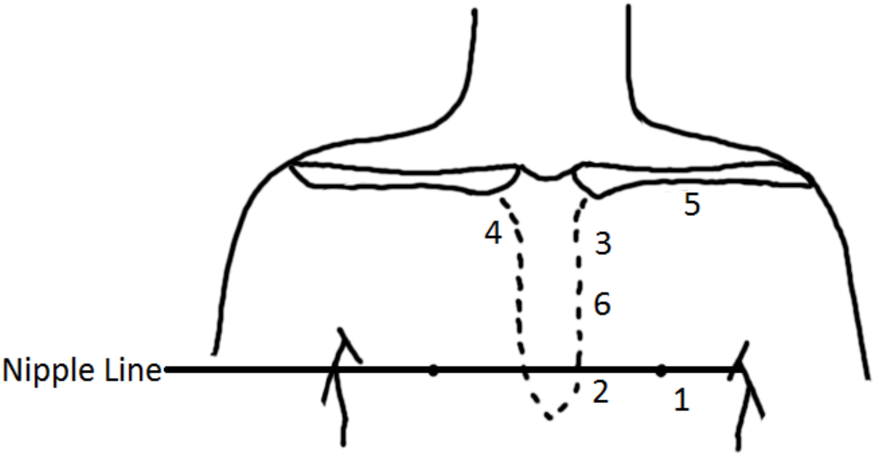


Figure 2
The position of auscultation areas
The 4 basic areas:
1. Apex. 2. Tricuspid area. 3. Pulmonary area. 4. Aortic area.
Special areas:
5. Infraclavicular area.
6. Second aortic area.

Blood Pressure:
Check the blood pressure using the appropriate cuff. The correct width of the BP cuff is 40% to 50% of the *circumference* rather

than the *length* of the limb on which the BP is being measured .Get blood pressure in the four limbs, normally the BP higher in lower limbs. If upper limb BP is more than lower limb by more than 15 mmHg COA is likely**.** BP is ideally measured for all children particularly those with risk factors for the metabolic syndrome. Reference values for BP in different ages are shown in Tables 3 and 4.

Table 3
Normal Blood Pressure Levels in Neonates and Children <5 years

Age	**BP: 50th % systolic/diastolic (mean)**	**BP: 95th % systolic/diastolic (mean)**
1-3 days	64/41 (50)	78/52 (62)
1 month- 2 years	95/58 (72)	110/71 (86)
2-5 years	101/57 (74)	115/68 (85)

Modified from Park MK, Menard SM: Normative oscillometric blood pressure values in the first 5 years in an office setting. Am J Dis Child 143:860, 1989.

Pulse Pressure:

Pulse pressure is the difference between systolic and diastolic BP. A wide pulse pressure is associated with conditions that decrease the diastolic pressure e.g. PDA and AR. A narrow pulse pressure is caused by a low systolic and high diastolic pressure, typically in pericardial effusion with tamponade.

Table 4
Normal Blood Pressure (systolic/diastolic) for Children 5-17 years

Age (yr)	**50th Percentile systolic/diastolic**	**95th Percentile systolic/diastolic**
5	104/58	118/76
6	106/59	120/67
7	107/59	121/77
8	108/60	123/78
9	110/61	124/78
10	110/61	125/79
11	113/62	127/79
12	115/62	129/79
13	118/62	132/79
14	120/62	134/79
15	122/62	136/79
16	123/62	137/80
17	123/62	137 /80

Modified from Park MK, Menard SW, Schoolfield J: Oscillometric blood pressure standards for children. Pediatr Cardiol 26:601–607, 2005.

Ambulatory BP monitoring:

BP reading at the clinic would not represent the true level as BP is changing with time and patient’s psychological conditions.Fear from physicians (white coat hypertension) had been reported in 7% of children while measuring their BP in the hospital setting.

Ambulatory monitoring can provide more accurate BP reading over 24 hours therefore day-night variability can be determined.

Home BP Monitoring:

It was found to be an effective way to overcome the problem of white coat hypertension. The readings are comparable to those of ambulatory BP monitoring.

Invasive BP Monitoring:

Patients in the intensive care set up often need invasive BP monitoring for central venous pressure (CVP) and arterial BP. CVP can be monitored through a central venous line inserted in the internal jugular (common), subclavian or femoral veins. The CVP indicates the cardiac filling pressure (preload) as well as the ventricular end-diastolic pressure.

Table 5

Interpretation and management of Abnormal CVP

Low CVP

Cause	**Action**
Dehydration	Increased fluid intake

High CVP

Fluid overload	Decrease fluid intake, diuretics
Cardiac tamponade	Confirm by Echo, pericardiocentesis
CHF (cardiomyopathy, VSD or MR)	Diuretics, treatment of CHF, inotropic drugs

Arterial pressure:

A radial, brachial or femoral artery catheter is inserted to monitor the arterial pressure. It should be correlated to the non-invasive BP to get a true idea of the patients BP status. Management of hypertension depends on the cause and the physicians preference for drugs. The most common cause of hypertension in the setting of cardiology is CoA. Betablockers (e.g. propranolol) is the drug of choice.

Systemic Hypotension (Circulatory Shock):

Table 6

Types and management of circulatory shock

Type	Action
Pre-cardiac (preload) Hypovolemic shock	Fluid replacement.
Cardiac CHF (cardiomyopathy, CHD)	Diuretics, Inotropics, inodilators, surgery
Post-cardiac (afterload) Septic shock (vasodilatation), Anaphylaxis	Inotropics, vasoconstricting agents (e.g.epinephrine and nor epinephrine)

Clinical Examination of *Children* with Heart Disease:

- The examination is more systematic but the *Golden Rules* still need to be applied. It is the same as examination of adults.
- Inspection is the same as in infants, in addition, inspect the patient's oral cavity to look for dental cares.
- Examine the hands: clubbing, cyanosis, pallor and signs of endocarditis:
  - Osler's nodes (tender, pea-sized red nodes at the ends of the fingers or toes).
  - Janeway lesions (small, painless, hemorrhagic areas on the palms or soles), splinter hemorrhages (linear hemorrhagic streaks beneath the nails) which are rarely seen in children.

- Roth's spots are oval, retinal hemorrhages with pale centers located near the optic disc occur in less than 5% of patients

- Examine the eyes for pallor and jaundice.
- Examination of the skin: petechiae on the skin and mucus membrane can be seen in endocarditis. Erythema marginatum is a major manifestation of acute rheumatic fever characterized by nonpruritic serpiginous or annular erythematous rashes are most prominent on the trunk and the inner proximal portions of the extremities. They have evanescent nature (disappear in cold and appear with hot weather). Other skin rashes as in systemic lupus and rheumatoid arthritis can be associated with cardiac involvement.
- Inspect the neck with the patient lying at 45 degrees to look for the jugular venous pressure. Prominent neck pulsation can be arterial (pulsatile) or venous (wavy and compressible). If the upper limit of the pulse cannot be visualised, let the patient sit up as the upper limit could be higher than the jaw. Prominent pulsations at the suprasternal notch indicates severe AR. Other signs of severe AR: Head nodding and very prominent ‘pistol shot’ femoral pulse
- Auscultate the lungs for basal crepitations.
- Examine the lower limbs for oedema.

Objective Assessment of CHF:

- The NYHA classification is helpful in older children to quantify the degree of CHF but it cannot be applied to infants.

- **The Ross Score**: is useful to quantify CHF in infants, this was proposed by Ross in 1992, it includes 11 variables which are : the amount of formula consumed per feeding, feeding time, history of diaphoresis or tachypnea, growth parameters, respiratory and heart rates, respiratory pattern, perfusion, presence of oedema, diastolic filling sounds, and hepatomegaly.
- We propose a simpler score that includes the heart and respiratory rates, weight and liver size, this score is more practical to quantify CHF in infants.

Table 7

A proposed CHF Score in infants (Sudan Heart Failure (SHF) Score)

Feature	Score
Heart Rate (beat/minute)	
100-120	1
>120-150	2
>150	3
Respiratory Rate	
30-40	1
>40-50	2
>50	3
Liver Span (cm)	
5 cm or less	1
5-7	2
>7	3
Weight for age	
Normal or one standard deviation (SD) below the mean	1
>1-2 SD below the mean	2
>2 SD below the mean	3

Degree of CHF	**Total score**
Mild CHF	4-6
Moderate CHF	>6-8
Severe CHF	> 8

Suggested Readings:

1. Park MK. Basic Tools in Routine Evaluation of Cardiac Patients. In: Pediatric cardiology for practitioners.5th ed. Philadelphia, Mosby, Elsevier2008.
2. van Beynum IM, Kapusta L, Bakker MK, den Heijer M, Blom HJ, de Walle HE. Protective effect of periconceptional folic acid supplements on the risk of congenital heart defects: a registry-based case-control study in the northern Netherlands. Eur Heart J. 2010 Feb;31:464-71.
3. Furusawa EA, Filho UD, Junior DM, Koch VH. Home and Ambulatory Blood Pressure to Identify White Coat and Masked Hypertension in the Pediatric Patient. Am J Hypertens.2011 (Pubmed)
4. Ross RD, Bollinger RO, Pinsky WW. Grading the severity of congestive heart failure in infants. Pediatr Cardiol 1992;13:72-5.
5. Mutaz J, Sulafa KM Ali. The correlation between clinical scores, echocardiographic scores and plasma natriuretic peptide in paediatric patients with heart failure (unpublished data)

Chapter 2

Investigations of Children with Heart Disease

2.1 Chest X-Ray and other Radiological Investigations

The value of CXR

1. **Heart size**
2. **Lung vascularity (plethora/oligemia)**
3. **Specific CXR appearance in CHD**
4. **Lung pathology associated with heart disease**

The Quality of the Film:

- Position of the patient: Antero-posterior, supine or tilted positions can lead to false cardiomegaly
- Penetration: over penetration can lead to underestimation of pulmonary vascularity and vice versa.
- Inspiratory vs. expiratory films: affect the heart size: expiratory films causing false cardiomegaly. This can result from shallow breathing (e.g neonates). Abdominal distension secondary to organomegaly or acsites can lead to poor inspiration.
- Labelling: very important as abnormal heart and visceral position can be present**.**

Cardiac Size and Shape:

- Note the cardiothoracic ratio (Figure 3) : in neonates the ratio could be up to 60% for the following reasons:

1. The thymic shadow is large and may overlap the heart.
2. The film is usually supine.
3. The film might be expiratory.
4. The lungs might not yet be fully inflated.

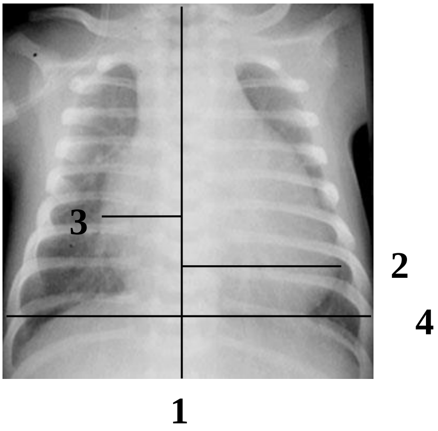


Figure3: Determination of the cardio-thoracic ratio (CTR): line 1: at the middle of the vertebral bodies, Line 2: maximum cardiac enlargement to the left, Line 3: maximum cardiac enlargement to the right, Line4: thoracic diameter. CTR= (2+3/4) X100. It is 50% where line 1and 4 cross.

Which chamber is enlarged? (Fig 4)

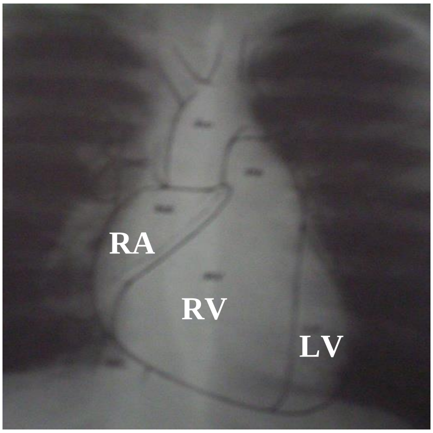


Figure 4
Antero-posterior CXR showing cardiac chamber positions

1. The RV is an anterior structure and the LV is posterior and left sided, LA is posterior and RA lies to the right occupying the right heart border in the antero-posterior film.
2. In the antero-posterior film the RA and LV enlargement can be evaluated directly while LA and RV can be evaluated *indirectly* as follows:
    - LA enlargement is evaluated by looking at the elevation of the left main bronchus, straight left heart border (Figures 5 and 7) or a double shadow behind the RA.
    - RV enlargement can be suspected when the apex is rounded and raised (typically in boot shape CXR).(Fig 6)
3. Note the size of the pulmonary artery and its main hilar branches

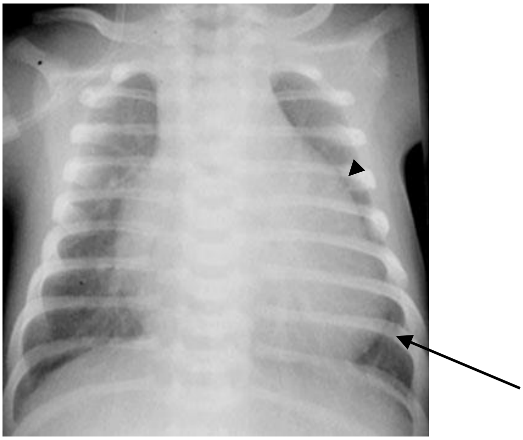


Figure 5

CXR of a patient with severe MR showing cardiomegaly : LV (arrow) and LA enlargement (arrow head) are evident. Pulmonary venous congestion is seen.

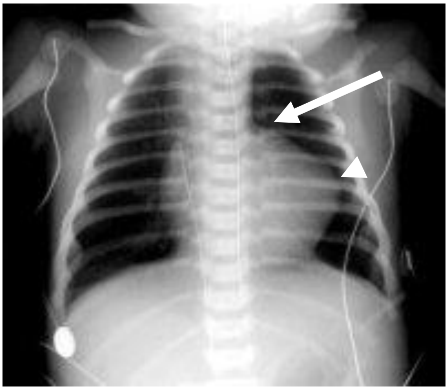


Figure 6

Chest Xray of a patient with TOF showing a ‘boot shape’ heart with rounded and raised apex (arrow) and concave main pulmonary artery segment (arrow head).(Note that the film is over-penetrated giving the lungs more oligemic appearance)

Lung plethora:

Evaluation of lung plethora is crucial in the diagnosis of paediatric heart disease; these are some important patterns for abnormal plethora:

1. **Increased pulmonary arterial markings:** seen as prominent central and peripheral pulmonary arteries which indicate large left to right shunts like VSD, ASD and PDA.(Figure7)
2. **Decreased pulmonary vascularity** which indicates lesions with decreased pulmonary blood flow like TOF and pulmonary atresia (Figure 6).
3. **Prominent pulmonary venous markings** seen as hazy/ fluffy interstitial shadows (pulmonary oedema) which occurs in lesions with increased left atrial/pulmonary venous pressure e.g MR and MS.(Figure 5)

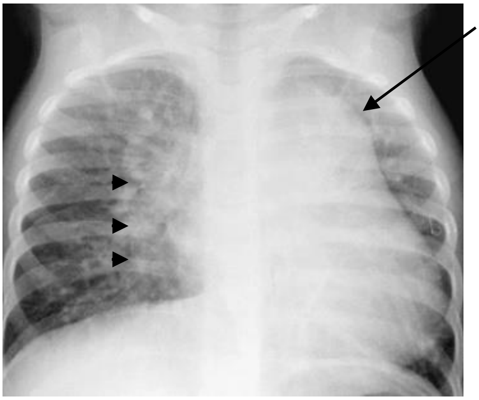


Figure 7

CXR of a patient with large VSD showing enlargement of LV, LA and main pulmonary artery (arrow) with prominent pulmonary arterial markings (arrow heads).

4.**Prominent central pulmonary arteries with peripheral oligemia:** indicate irreversible pulmonary vascular obstructive disease (Eisenmenger's syndrome).(Fig 8)

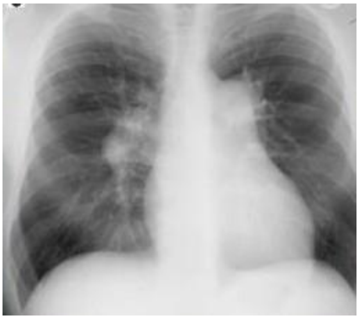


Figure 8

CXR of a 15 year old patient with Eisenmenger’s syndrome showing prominent central pulmonary arteries with sudden oligemia (pruning) peripherally.

Sidedness of the heart, aorta and abdominal organs (Situs):

1. Look for the position of the apex: normal (levocardia), to the right (dextrocardia) or in the middle (mesocardia).Dextroposition referes to the shift of the

heart to the right secondary to abnormalities in the lung.e.g hypoplasia of the right lung.

2. If the trachea is normally shifted to the right the aortic arch is left sided (normal). Right sided aortic arch causes the trachea to be central or shifted to the left.
3. Look for the gastric bubble and the liver shadow, a left sided stomach and right sided liver indicate normal abdominal situs (situs solitus: normal arrangement). Abdominal situs inversus is present when the liver lies on the left and stomach on the right. Situs ambiguous is usually associated with a central liver (Figure 85)

Lung pathology:

1. It is often difficult to differentiate radiologicaly primary lung pathology and that related to the effect of large left to right shunt, clinical correlation is important.
2. Pneumonia is a common association in patients with large left to right shunts.
3. Left lower lobe collapse due to compression of the left bronchus by the enlarged LA can occur in patients with large VSD or PDA.
4. Hyperinflation is common in left to right shunts, it is due to air trapping caused by valve-effect mechanism secondary to compression of the bronchi by the enlarged pulmonary arteries.
5. **Specific lung Syndromes in CHD:**

I.**Scimitar syndrome**: hypoplasia of the right lung with sequestration of the left lower lobe which is supplied from the aorta and has anomalous pulmonary venous drainage to the inferior vena cava.(Figure9)

II. **TOF with absent pulmonary valve syndrome**: CXR shows huge pulmonary arteries which can be associated with lung collapse/infiltrations that makes the CXR appearance more suggestive of respiratory disease. (Figure 10)

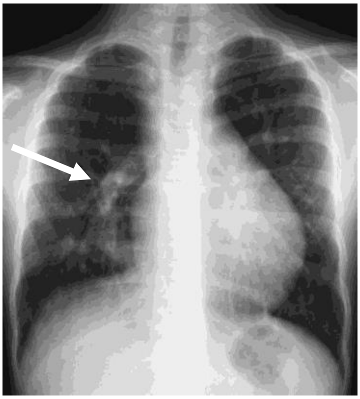


Figure 9
CXR of a patient with Scimitar syndrome showing a cresentic shadow in the right lung (arrow) due to the abnormal pulmonary venous drainage of the sequestered segment to the inferior vena cava.

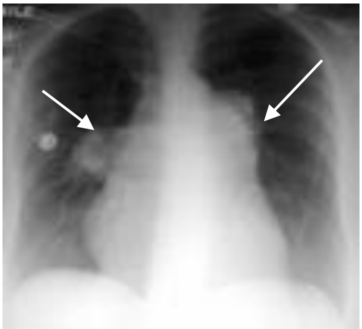


Figure 10
CXR of a patient with TOF-Absent pulmonary valve syndrome showing prominent right and left pulmonary arteries. (arrows)

Cont. CXR Interpretation:
Skeletal abnormalities:

1. Rib notching: In older children/adults with CoA rib notching is due to the presence of collateral arteries.(Fig 11)
2. Congenital skeletal anomalies in syndromes associated with CHD: e.g.TAR: **T**hrombocytopenia with **A**bsent **R**adius syndrome, butterfly vertebrae in Alagille's syndrome.

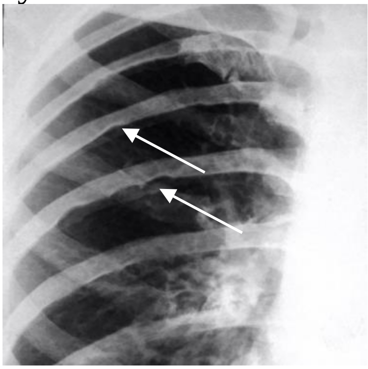


Figure 11:
Rib notching (arrows)

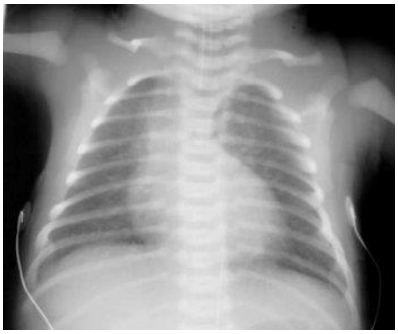


Figure 12
CXR of a patient with TGA showing "egg on side appearance"

Table 8:
Classical CXR appearance in CHD

X ray Appearance	**Diagnosis**
Boot shaped: (Fig 6) • Small heart with round apex. • Concave pulmonary artery segment. • Oligemic lungs	TOF and TOF - Like conditions
Egg on side: (Fig 12) • Narrow heart pedicle/base. • Oval, big heart. • Plethoric lungs	TGA
Snowman appearance: (Fig 13) Large supracardiac shadow	TAPVD (supracardiac)

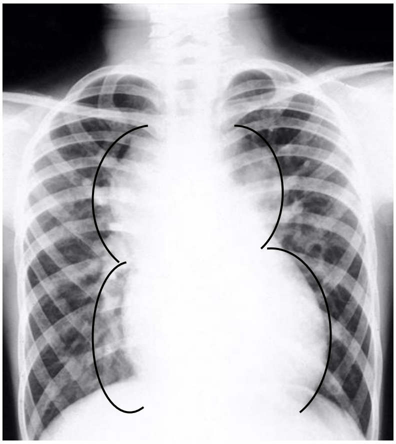


Figure 13

CXR of a patient with supracardiac TAPVD showing a “figure of 8” (snowman) appearance

Other radiological investigations:

Cardiac MRI

Cardiac MRI has rapidly developed and became increasingly utilized for evaluation of patients with CHD. The advantages of cardiovascular MR include a large field of view, unlimited choice of imaging planes and low operator-dependency. The most important indications are:

1. Delineation of the anatomy of pulmonary arteries and veins, aortic arch and its branches.(Figure 14)
2. Functional assessment of the right ventricle.
3. Quantification of valve regurgitation.

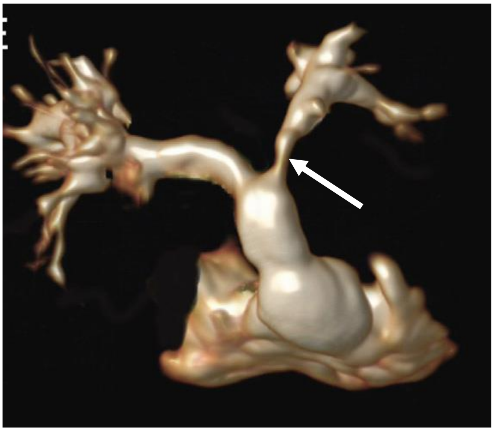


Figure 14
Gadolinium-enhanced MR angiogram of a patient with repaired TOF showing severe proximal left pulmonary artery stenosis (arrow)

Cardiac CT:

The development of spiral and subsequently multi-detector computed tomography (MDCT) enables the acquisition of data during a single breath-hold and during the first pass of a contrast bolus, so that images can be reconstructed in any two-dimensional or 3-dimensional planes. A major advantage of the recent advances in CT technology over MR is the very rapid acquisition time. It can delineate the anatomy of PA branches, aortic arch and vascular rings (Figure 15). These non invasive investigations had largely replaced cardiac cath for the diagnosis of CHD.

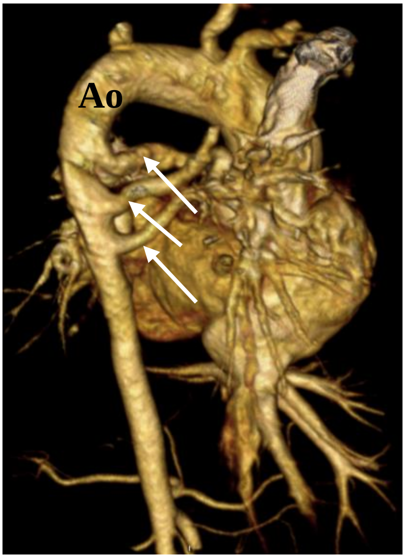


Figure 15
64-Multi slice CT image in a patient with pulmonary atresia/VSD showing multiple aorto pulmonary collaterals (arrows) form the desending aorta (Ao)

2.2 ECG Interpretation

The limb leads (leads I, II, III, aVR, aVL, and aVF) provide information about the frontal projection (reflecting superior-inferior and right-to-left forces), and the precordial leads (leads V1 through V6, V3R, and V4R) provide information about the horizontal plane that reflects forces that are right to left and anterior-posterior.

Types of ECG:

1. 12 lead ECG: The regular ECG, if special paediatric leads (V3R and V4R) are used it will be 15 leads.(Figure16)
2. Exercise ECG: Done while the patient is exercising (Treadmill-Stress ECG) for patients at suitable age (usually >7 years of age). It is indicated when symptoms are related to exertion like AS where echo is not conclusive. Also for exercise related rhythm disorders. It has to be requested and attended by the paediatric cardiologist.
3. Holter monitor: ambulatory ECG where 2-4 leads are connected for 24 hours or more. It is indicated in patients with arrhythmias when the symptoms are frequent.
4. Event monitor: A device that is connected to the chest and activated by the patient or parents when symptoms occur. The recorded ECG is transmitted by telephone to the cardiac centre.
5. Trans esophageal ECG using a small lead placed in the esophagus can help to delineate the arrhythmia and can also be used to for pacing.
6. Implantable loop recorders: devices implanted subcutaneously and activated by the patient when

symptoms occur. It is indicated for patients with infrequent but serious symptoms when other recording methods fail. The records can be analyzed *in vivo* to determine the abnormal rhythm.(Figure 17)

7. Invasive electrophysiological studies: done through cardiac cath to map the electrical activities (diagnostic) and ablate abnormal foci of discharge like accessory pathways (interventional).

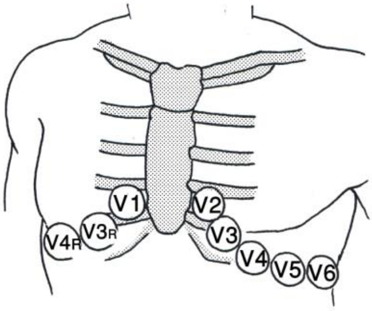


Figure 16
Precordial lead position showing the ‘special’ pediatric leads V3R and V4R in addition to the standard chest leads.

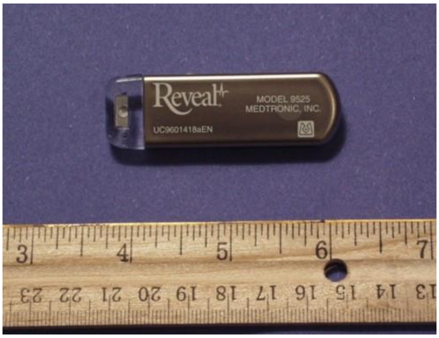


Figure 17
Implantable loop recorder

The differences between paediatric and adult ECG:

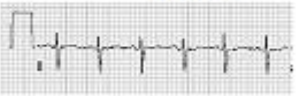
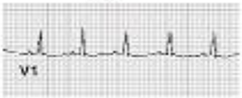
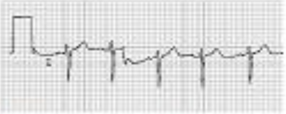
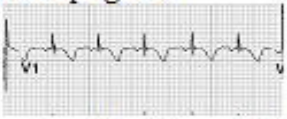
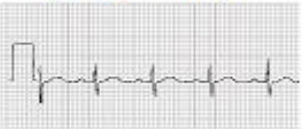
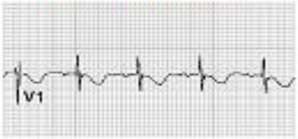
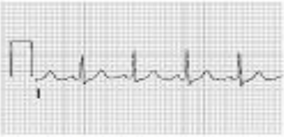
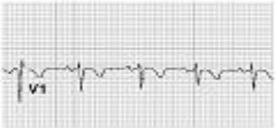
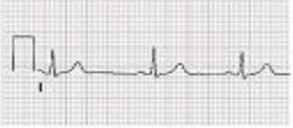
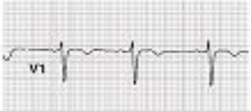
1. *Right ventricular dominance*: the RV is dominant from birth up to around 2 years then the LV takes over. RV dominance is best evaluated by the R/S ratio in V1 which is shown in ‘milestones of ECG’ below.
2. *Lead position*: due to RV dominance additional paediatric leads are used, namely V3R and V4R (Fig 16) up to around 8 years. These reflect RV forces.
3. *Right QRS axis*: also related to RV dominance. The axis shifts to the left gradually after infancy.
4. *T wave axis*: the T wave is flat or upright at birth then *inverts* from the age of 72 hours up to around 8 years. Upright T wave in infancy and childhood is a sensitive indicator of RVH.

ECG Milestones:

ECG milestones are summerised in table 9. Most of these changes occur in leads I and V1 which sho :

1. Shift of QRS from negative to positive in lead 1
2. Inverted T in V1
3. Increasing depth of S and decreasing voltage of R in V1

Table 9
The milestones of ECG:

Age	*Lead I*	*V1*
0-3 days	 Negative voltage (Rt QRS axis)	 Pure R, no S Flat/upright T
3 days – 6 month	 Negative voltage (Rt QRS axis)	 Dominant R with small S Well-inverted T wave
1 – 2 year	 equivocal voltage (QRS axis shifting to the left)	 Larger S wave, R/S =1 Well inverted T wave
2 years- 8 y	 Positive voltage (Left QRS axis)	 Deep S, R/S ratio<1
8 years – adults	 Left QRS axis	 Deeper S, Slower heart rate

ECG Interpretation

Systematic evaluation with reference to normal values (Table 5):

1.The Rate:

- Count the big squares between two QRS complexes and divide 300 by the no of big squares, this is the *ventricular rate.*
- Count the big squares between two P waves and divide 300 by the no of big squares, this is the *atrial rate.*
- Atrial and ventricular rates should be the same if the patient is in normal sinus rhythm.

3. **The Rhythm:**
   - Check the P wave morphology and axis: if each p is followed by QRS and the p wave axis is normal (upright p in leads I,II,III and aVF and inverted P in lead AVR) the *rhythm is sinus*.

4. **Atrial Enlargement:**

- RA enlargement: P wave height >2.5 mm (2.5 small squares)
- LA enlargement: P wave width > 0.08 secs (2 small squares), bifid P wave.

Ventricular Hypertrophy:

Criteria for RVH:

- Upright T wave in the right precordial leads 5 days- 8years of age
- Voltage criteria: R in V1 > 20mm at any age.

- Pure R in V1 after neonatal period.
- R/S ratio > 1 after 24 months.
- qR pattern in V1
- Volume Overload pattern:
  - Rsr' pattern in V1 often with normally inverted T wave
  - qR pattern in V1
- Pressure overload pattern:
  - Tall R in V1 (voltage criteria) with upright T wave.

Criteria for LVH:

- Tall R in V6 (> 25 mm)
- Deep S in V1 (> 25 mm)
- Summation of S in V1 and R in V6 > 35 mm

Criteria for Biventricular Hypertrophy:

- Summation of R and S in mid precordial leads (V2-5) > 55 mm (called the Katz-Wachtel phenomenon)
- Criteria for both RVH and LVH

6. Intervals and Durations:

- PR interval: short PR is seen in WPW and glycogen storage disease. Long PR (first degree AV block) is seen in acute rheumatic fever and AVSD.
- QRS duration: wide QRS is seen in ventricular arrhythmia, post cardiac surgery RBBB.
- QT interval: Calculation of corrected QT (QTc) is very important because of the association of long QTc with sudden cardiac death (long QT syndrome-see scenario14).

Correction should be done using the Bazzet's Formula: QTc = measured QT/√preceding RR interval (msec)

NB: If the heart rate is 60 beats/minute or less there is no need for calculation of corrected QTc.

7. T wave and ST segment:

- Elevated ST segment followed by T wave inversion: pericarditis.
- Inverted T wave in V6: LV strain pattern.
- Myocardial infarction: rare; can be due to anomalous left coronary artery from pulmonary artery or Kawasaki disease. Characterized by: pathological q wave, ST segment elevation and T wave inversion.

8. QRS Axis:

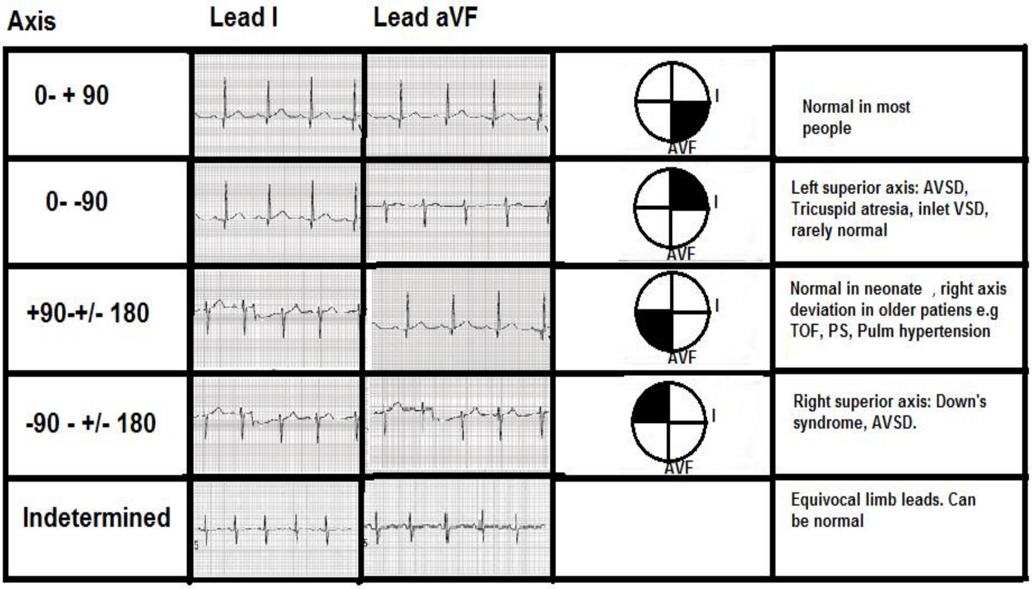
- Calculate using the hexa-axial reference system and diagram shown in Figure 18.

ECG in Electrolyte Disturbances:

- Hypocalcemia prolongs the ST segment, resulting in a prolonged QTc.
  Conversely, hypercalcemia shortens the ST and QTc.
- Hypokalemia produces prolonged PR interval, flattening of T waves, and prominent U waves.
- Hyperkalemia is a serious condition because it can result in fatal ventricular arrhythmia. Therefore care has to be taken when admistering potassium and potassium retaining drugs. (Figure 19)

Table 10: Normal ECG values according to age.

Age	HR bpm	QRS axis degrees	PR interval seconds	QRS interval seconds	R in V1 mm	S in V1 mm	R in V6 mm	S in V6 mm
1st week	90-160	60-180	0.08-0.15	0.03-0.08	5-26	0-23	0-12	0-10
1-3wks	100-180	45-160	0.08-0.15	0.03-0.08	3-21	0-16	2-16	0-10
1-2 mo	120-180	30-135	0.08-0.15	0.03-0.08	3-18	0-15	5-21	0-10
3-5 mo	105-185	0-135	0.08-0.15	0.03-0.08	3-20	0-15	6-22	0-10
6-11 mo	110-170	0-135	0.07-0.16	0.03-0.08	2-20	0.5-20	6-23	0-7
1-2 yr	90-165	0-110	0.08-0.16	0.03-0.08	2-18	0.5-21	6-23	0-7
3-4 yr	70-140	0-110	0.09-0.17	0.04-0.08	1-18	0.5-21	4-24	0-5
5-7 yr	65-140	0-110	0.09-0.17	0.04-0.08	0.5-14	0.5-24	4-26	0-4
8-11 yr	60-130	-15-110	0.09-0.17	0.04-0.09	0-14	0.5-25	4-25	0-4
12-15 yr	65-130	-15-110	0.09-0.18	0.04-0.09	0-14	0.5-21	4-25	0-4
> 16 yr	50-120	-15-110	0.12-0.20	0.05-0.10	0-14	0.5-23	4-21	0-4



Axis	Lead I	Lead aVF		
0- + 90				Normal in most people
0- -90				Left superior axis: AVSD, Tricuspid atresia, inlet VSD, rarely normal
+90-+/- 180				Normal in neonate , right axis deviation in older patiens e.g TOF, PS, Pulm hypertension
-90 - +/- 180				Right superior axis: Down's syndrome, AVSD.
Indetermined				Equivocal limb leads. Can be normal

Figure 18
QRS axis position and the differential diagnosis of the five possible positions.

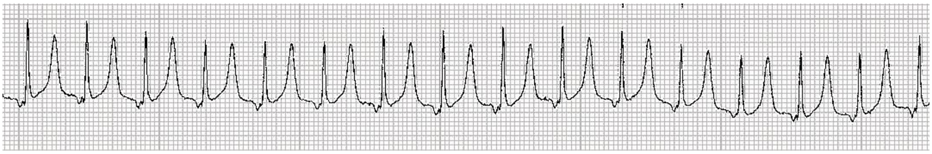


Figure 19-A:
Peak tall P wave in a patient with serum K 7.4 meq/l

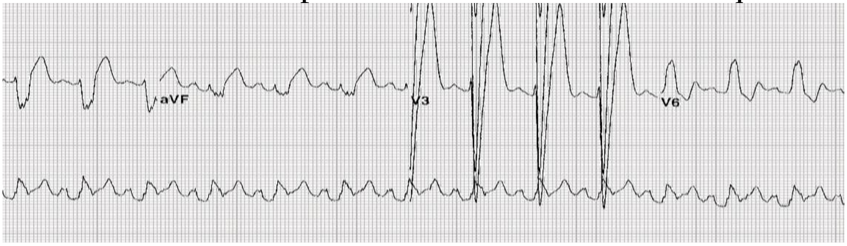


Figure 19-B
Ventricular tachycardia (wide QRS) in a patient with serum potassium of 8 meq/l

2.3 Basic Echocardiography

- Ultrasound (U/S) is sound waves with a frequency of > 20 000 Hz (cycles/sec) therefore it cannot be heard by the human ear.
- Echo is an advanced type of ultrasound modified to penetrate the thoracic tissues to delineate the structure of the heart and vessels.
- Echo can reliably diagnose structural anomalies (congenital, valvular or mass lesions) as well as functional assessment of hemodynamics (pressures, shunts and volumes).

How is the image produced?

- Ultrasound transducers with different frequencies (2-12 megahertz) use a piezoelectric crystal (that can change electric current into ultrasound) to generate and receive ultrasound waves.
- The transducer transmits bursts of ultrasound and then switches to the receive mode to await the reflected ultrasound signals from the intracardiac acoustic interfaces. These images are then displayed on the screen.

What are the current available modes of Echo and Doppler?

- 2-Dimentional echo: is a live 2-D image showing different structures cut at standard angles (standard echo windows, see below)
- 3-Dimentional echo: Using special transducers the image is processed by computer programmes to generate a 3-D image. 3-D echo has increased the diagnostic accuracy of echo for structural heart disease.

- M-Mode: single plane static image showing lines representing anatomical structures, used mainly to measure ventricular function and chamber size. (Fig 20)
- Doppler ultrasound: Is based on recording blood velocity. The direction of the blood indicates the colour; red colour indicates blood coming towards the transducer and blue colour indicates the opposite. Pulse and continuous wave Doppler measure the velocity in meters per second displayed as a graph (Fig 21). It is very useful to quantify gradients across valves, vessels and chambers. Gradients are calculated using the modified Bernoulli equation (gradient = 4x velocity ^{2}). Colour Doppler is a semi-quantitative method for valvular dysfunction as well as visualization of shunts. To obtain the most accurate prediction of the peak pressure gradient, the Doppler beam should be aligned parallel to the jet flow, the peak velocity of the jet should be recorded from different transducer positions, and the highest velocity should be taken.
- Trans-oesophageal echo: uses a small transducer mounted on an endoscope. It has advantage over the conventional trans-thoracic echo especially in patients with poor windows (e.g adults), monitoring of surgical or cardiac catheterization procedures and visualization of prosthetic valves.
- Foetal echo: foetuses suspected to have CHD on antenatal U/S are referred to a paediatric cardiologist for foetal echo. This procedure needs an experienced operator. It has an important role in early detection of CHD. In certain centres some interventions can be done in utero to foetuses with CHD like balloon dilatation of aortic stenosis.
- Other modalities: intravascular ultrasound is done by microtransducer mounted on a cardiac catheter and used through the femoral vein.

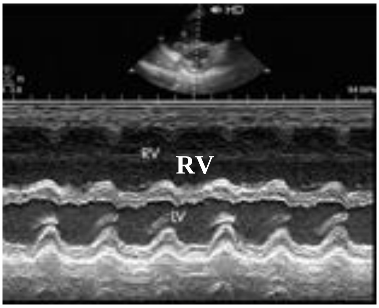


Figure 20-A
M-Mode echo showing a dilated RV.

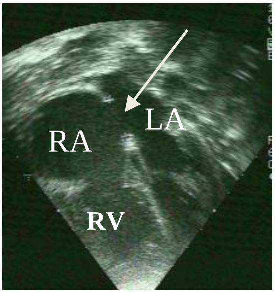


Figure 20-B

4 Chamber view of the same patient showing a large ASD (arrow)

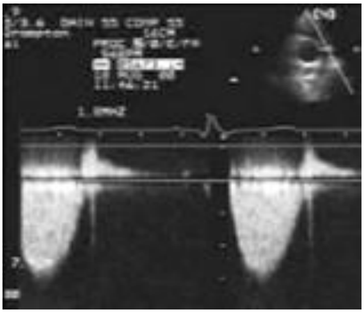


Figure 21
Continuous wave Doppler from a patient with mild pulmonary stenosis showing a peak velocity of 3.2 meters/second corresponding to a peak gradient of 40 mmHg.

What is a good Echo Study?

- Complete and systematic using the standard echo windows (Figure 22).
- Sequential analysis: Systematic search of cardiac anatomy starting from abdominal and atrial situs, atrial connections, atrio-ventricular and ventriculo-arterial connections and

systemic and pulmonary veins. Simultaneously, the operator searches for associated CHD which often occur in clusters.

- The patient is comfortable and quite (infants often need sedation)
- Answers questions raised by the requesting physician and gives all information needed by the surgeon according to the intended surgical procedure.
- Quantitative data with reference values
- Comprehensive report using standard terminology and nomenclature.

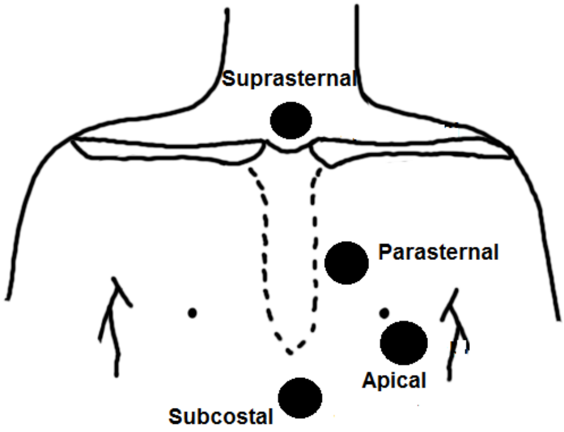


Figure 22
The Standard Echo Windows

Echocardiographic Views:

1. Subcostal window: Figure 23,24 and 25

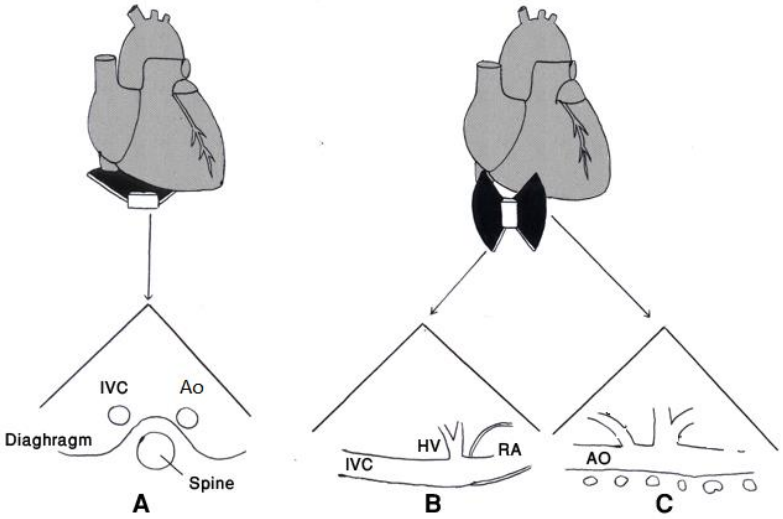


Figure 23:
Situs view: A: Short axis view , B: long axis view to the right. C: Long axis view to the left. (Ao: aorta, HV: Hepatic vein, IVC: inferior vena cava).

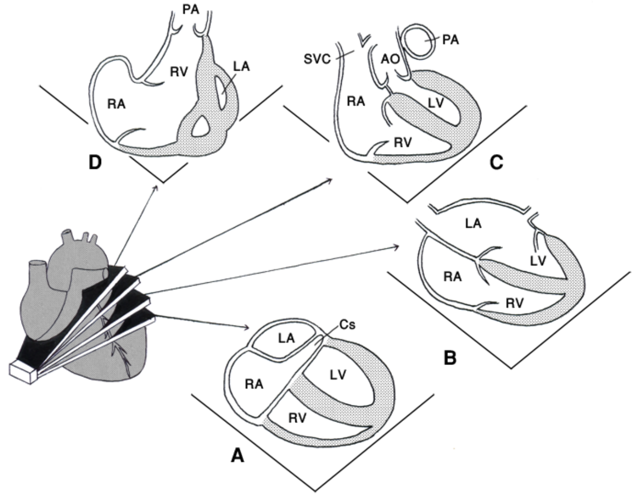


Figure 24:
Subcostal long axis view: A: coronary sinus (CS)view, B: 4-chamber view, C:LV outflow tract view,D: RV outflow tract view. (SVC:Superior vena cava)

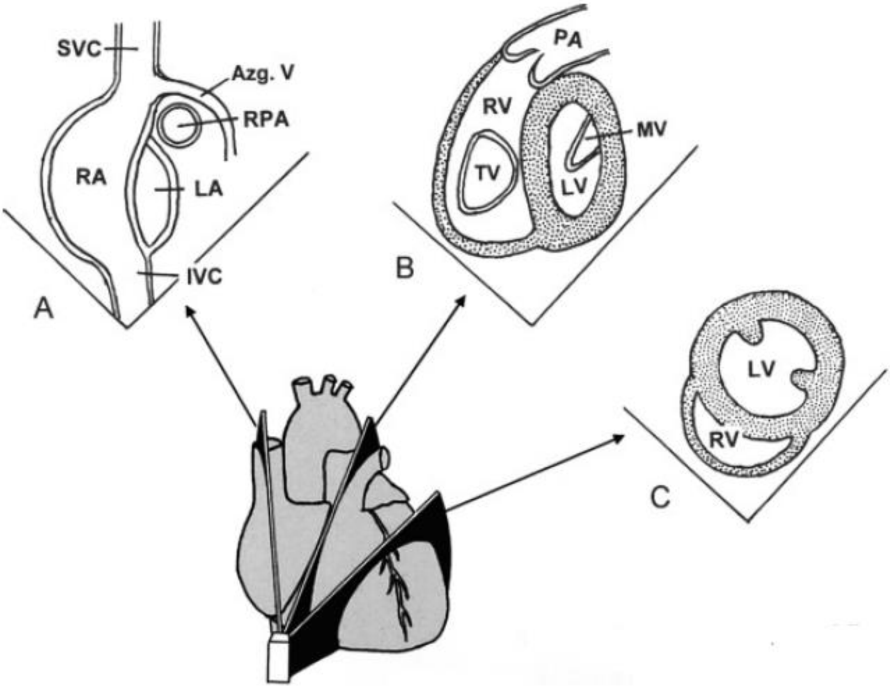


Figure 25:

Subcostal short axis view: A: Bicaval view, B: RV outflow tract view, C: short axis view. Azg v: azygous vein, SVC: superior vena cava, IVC: inferior vena cava.

2. Parasternal window: (Figure 26 and 27)

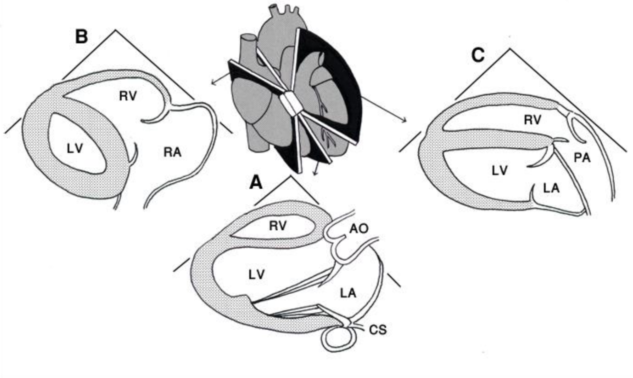


Figure 26 (above):
Parasternal long axis view: A: Standard view, B: tricuspid valve view, C: pulmonary valve view. (CS: Coronary sinus)

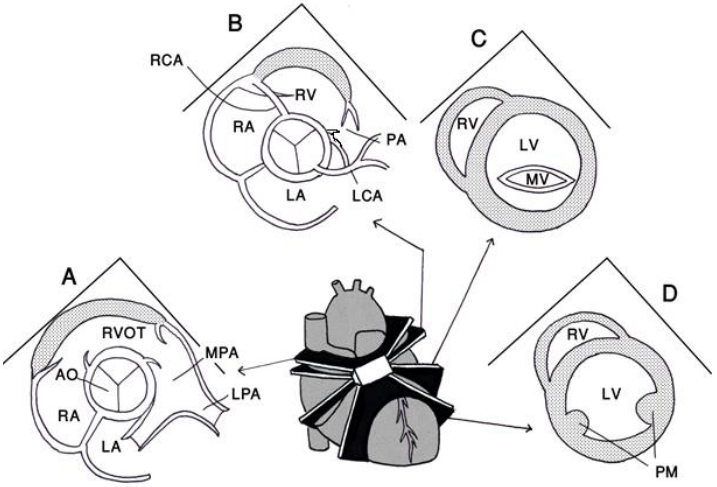


Figure 27:
Parasternal short axis view: A: PA branches view, B: Base of the heart view, C: Mitral valve view, D: Papillary muscle view.RCA: Right coronary artery, LCA: Left coronary artery.LPA and RPA: right and left PA.

3. Apical window: (Figure 28)

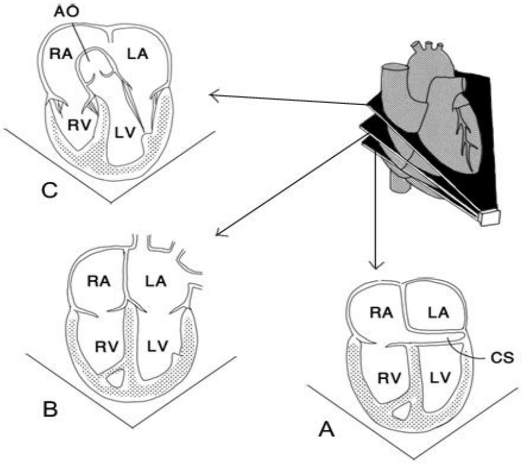


Figure 28:
Apical 4 chamber view: A: Coronary sinus (CS) view, B: AV valves view, C: 4-chmber with aorta view.

4. Suprasternal window: (Figure 29 and 30)

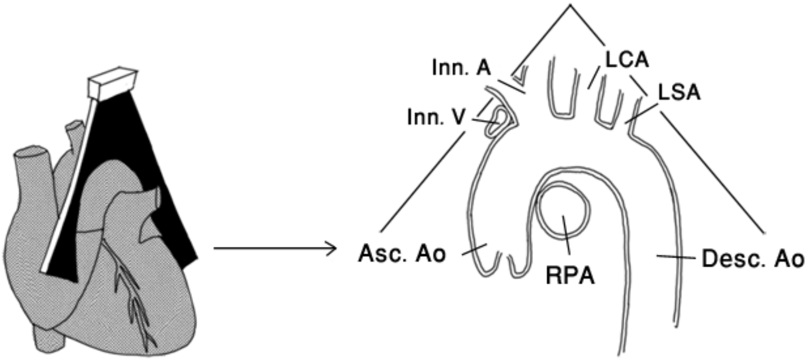


Figure 29:
Suprasternal long axis view showing Inn.A: innominate artery, LCA: Left carotid artery, LSA: Left subclavian artery, RPA: right PA.

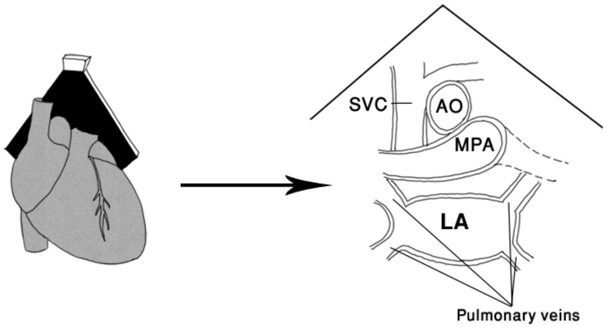


Figure 30:
Suprasternal short axis (Crab) view. MPA: main PA, SVC: superior vena cava.

3.4 Cardiac Catheterization

Cardiac cath refers to the use of catheters inserted through the large veins, arteries or both to study cardiac structure and hemodynamics (diagnostic cath) or treat cardiac diseases (therapeutic/ interventional cath).The cardiac cath laboratory consists of XR fluoroscopy tubes (anteroposterior and lateral in bi plane laboratories) and invasive manometers connected to the patients' vessels to measure pressures in different chambers. Blood samples are taken to measure oxygen saturation and blood gas analysis. Contrast injections done in the heart chambers and vessels taken in different projections help to delineate the anatomy.

Indications:

Diagnostic:

1. Delineate anatomical structures: certain parts of the heart and vessels cannot be well seen by echo like peripheral pulmonary branches and pulmonary veins. This is done

through angiography in the heart chambers and vessels using different projections.

2. Hemodynamic studies: measurement of pressures at different chambers most commonly pulmonary artery pressure, shunt quantification and pulmonary resistance calculation. This is done by recording the pressure and oxygen saturation in different chambers.

i. Left to right shunt (QP/QS)=
Aortic saturation- Systemic vein saturation
÷ Pulmonary vein saturation -Pulmonary artery saturation

ii. Cardiac output calculation : the Fick principle is used to measure the systemic (QS) and pulmonary(QP) blood flow

$$QP = VO2/ CPV - CPA$$
$$QS = VO2/ CAO - CMV$$

(where flows are in L/minute, VO_2 is oxygen consumption (mL/minute), C is oxygen content (mL/L) at various positions, PV is pulmonary vein, PA is pulmonary artery, AO is aorta, and MV is mixed systemic venous blood (superior vena cava or right atrium).

The normal systemic flow or pulmonary flow in the absence of a shunt is 3.1 ± 0.4 L/minute/m^2 (i.e., cardiac index). An assumed oxygen consumption of 150 to 160 mL/min/m^2 is used for older infants and children. For infants younger than 2 to 3 weeks, 120 to 130 mL/min/m^2 may be used.

iii. Pulmonary vascular resistance calculation:

$$PVR = \text{Mean PAP} - \text{Mean LAP}/ QP$$

This is done while the patient is breathing room air then repeated on 100% oxygen and/or other pulmonary vasodilators like nitric oxide. If resistance is more than 10 Wood's Units and did not drop on oxygen the patient is considered to have irreversible pulmonary vascular obstructive disease (Eisenmengers' syndrome) (see scenario 5)

Interventional Cath:

1. Closure of heart defects:

Rapid advances in this field resulted in availability of high quality occlusion devices for PDA, ASD and VSD. Other vessels like arterial or venous collaterals and fistulae can also be closed. PDA catheter closure is safe and reliable with the use of devices like Amplatzer Duct Occluder (Fig 31). ASD occlusion needs selection of central ASD secundum defects with good rims to hold the device. With this type of ASD the procedure is safe with high success rate.(Figure 77). Muscular VSDs can be closed reliably and safely in older patients, however closure of the more common perimembranous VSDs still not optimal with the risks of heart block and AR. The device technology is rapidly advancing and is likely to overcome these problems.

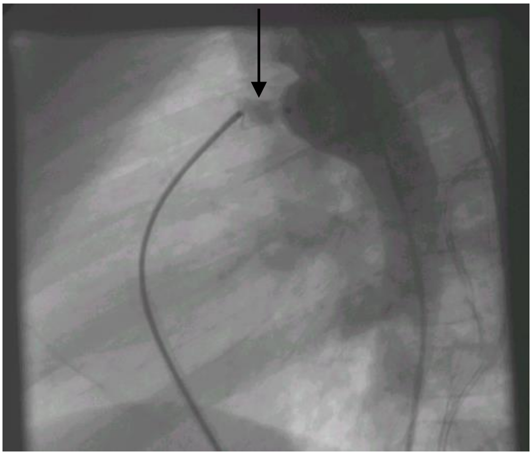


Figure 31

Angiographic lateral view aortogram showing a PDA device (arrow) well positioned and not yet released.

2.Dilatation of Valves:

Pulmonary valve dilatation (Transcatheter pulmonary valvuloplasty) is one of the earliest interventions. Most stenotic valves can be dilated except when the leaflets are dysplastic or the annulus hypoplastic as in patients with Noonan's syndrome.

Dilatation of the aortic valves requires careful selection of valves with no/minimal regurgitation and a balloon that is 90% of the valve annulus.

Dilatation of rheumatic mitral stenosis can be done reliably with careful selection of pliable leaflets with minimal regurgitation.

3. Balloon atrial septostomy:

This procedure was initially descried by Rashkind in 1962 . It is done for palliation of patients who need an atrial septal communication like TGA with intact ventricular septum. Other conditions that may need this procedure are TAPVD and Tricuspid atresia with restrictive ASD. It is usually done for neonates in the first few days of life when the fossa ovalis is patent. It can either be done through the umbilical or femoral vein access.

4.Coarctation stenting and balloon dilatation:

Native coarctation in infants and young children is treated surgically, however 20% of patients usually develop recoarctation which is best managed by balloon dilatation and/or stenting. In older children and adults stenting can be done as a primary procedure.

5.Transcatheter valve replacement: recently valves bound to stents could safely be placed into the pulmonary and aortic position using transcatheter techniques.

Potential complications of cardiac cath:

Diagnostic cath carries a low risk of complications related to catheter manipulations like transient arrhythmias or bleeding from puncture sites. More serious complications like perforation of heart vessels and hemopericardium are rare. Interventional cath can be associated with device embolization or malposition . Retrieval of the device can be attempted using special catheters but immediate surgical removal may be required.

Suggested Readings:

1. .Park MK, Guntheroth WG: *How to Read Pediatric ECGs*, 4th ed. Philadelphia, Mosby, 2006.

2. Snider AR, Serwer GA, Ritter SB: *Echocardiography in Pediatric Heart Disease*, 2nd ed. St.Louis, Mosby, 1997

3. Rajesh Puranik, Vivek Muthurangu,David S. Celermajer,Andrew M. Taylor. Congenital Heart Disease and Multi-modality Imaging. Heart, Lung and Circulation 2010;19:133–144.

4.Al Dhahri KN, Potts JE, Chiu CC, Hamilton RM, Sanatani S. Are implantable loop recorders useful in detecting arrhythmias in children with unexplained syncope? Pacing Clin Electrophysiol. 2009 ;32:1422-7.

5.Huhta JC, Gutgesell HP, Murphy Jr DJ, et al: Segmental analysis of congenital heart disease. *Dynamic Cardiovasc Imaging* 1987; 1:117-125.

6.Hu HB, Jiang SL, Xu ZY, Huang LJ et al. Outcome of interventional therapy of perimembranous ventricular septal defects.Zhonghua Xin Xue

Guan Bing Za Zhi (Chinese Journal of Cardiovascular Disease). 2009 ;37:618-21.

7.Waldhausen JA, Boruchow I, Miller WW,Rashkind WJ.Transposition of the great arteries with ventricular septal defect. Palliation by atrial septostomy and pulmonary artery banding. Circulation. 1969;39(Suppl 1):I215-21.

8.Kan JSz, White RI Jr, Mitchell SE, Gardner TJ2.Percutaneous balloon valvuloplasty: a new method for treating congenital pulmonary. N Engl J Med.1982;307:540-2.

9.Wilson ADRao PS, Galal O, Patnana M, Buck SH3.Results of three to 10 year follow up of balloon dilatation of the pulmonary valve. Heart.1998;80:591-5.

10.Atiq M, Aslam N, Kazmi KA. Transcatheter closure of small-to-large patent ductus arteriosus with different devices: queries and challenges. J Invasive Cardiol. 2007;19:295-8

11.Carminati M, Butera G, Chessa M, De Giovanni J. Transcatheter closure of congenital ventricular septal defects: results of the European Registry Eur Heart J. 2007;28:2361-8.

12.Frigiola A, Nordmeyer J, Bonhoeffer P.Percutaneous pulmonary valve replacement. Coron Artery Dis. 2009;20:189-91.

13.Patel JH, Mathew ST, Hennebry TA.Transcatheter aortic valve replacement: a potential option for the nonsurgical patient. Clin Cardiol. 2009 ;32:296-301.

Chapter 3

Approach to Common Presentations of Paediatric Heart Disease: Case Scenarios

General Approach:

Communication with the patient and family:

- Greet the patient and his parents, introduce yourself and let them sit comfortably. Take permission to examine the patient.
- Never take a baby from his mother! The best place to examine infants is their mothers lap while being breast fed. Stand at a distance of 1-1.5 meters from the patient to inspect.
- Talk to your patient gently while you are looking.
- Give the parents consideration and attention while examining their child.
- Listen to parent’s requests regarding the patient's position, needs and preferences.
- Always have a toy, candy or small gift for your patient

Inspection of the Patient:

Classify your patient according to the following:

1. Respiratory Distress.
2. Cyanosis/desaturation and clubbing.
3. The presence of wasting (failure to thrive).
4. Dysmorphic features.
5. The age.

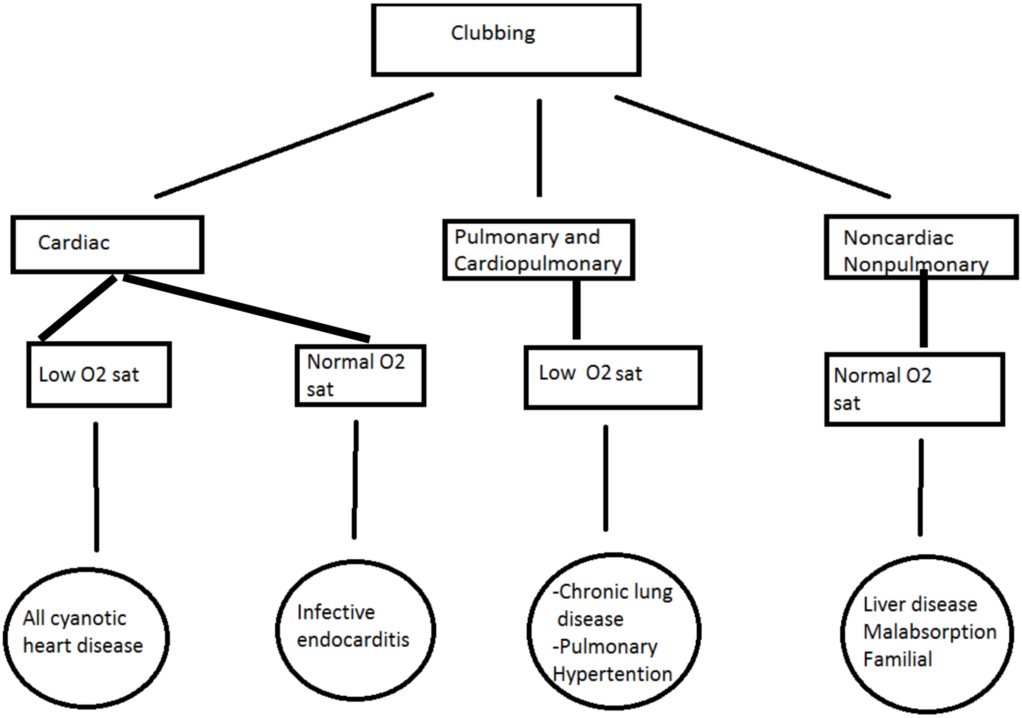
- Respiratory distress: in the presence of wasting and chest deformity indicates **chronic heart failure** or chronic respiratory disease.
- Respiratory distress *without wasting* can indicates :

- Acute heart failure (myocarditis)
- Cyanotic spell(in cyanotic patients)
- Acute respiratory disease

- At this stage peripheral cyanosis can help to indicate central cyanosis.

The vital questions to get a diagnosis by inspection:

- Is this a congenital or an acquired heart disease?
- Is it associated with normal or abnormal saturation: these terms are more accurate than the terms 'cyanotic' and 'acyanotic' as cyanosis can easily be missed clinically.
- Is it associated with heart failure or not?



Respiratory distress and failure to thrive are the most constant signs of chronic heart failure in infants

CHD are better classified as: CHD with normal O2sat and CHD with low O2 sat *rather than* cyanotic and acyanotic

The Chronological order of presentation in CHD:

Day 1-7
- Duct dependant conditions: TGA, CoA, pulmonary atresia, HLHS.
- Common mixing lesions like truncus arteriosus, tricuspid atresia.
- Obstructed TAPVD

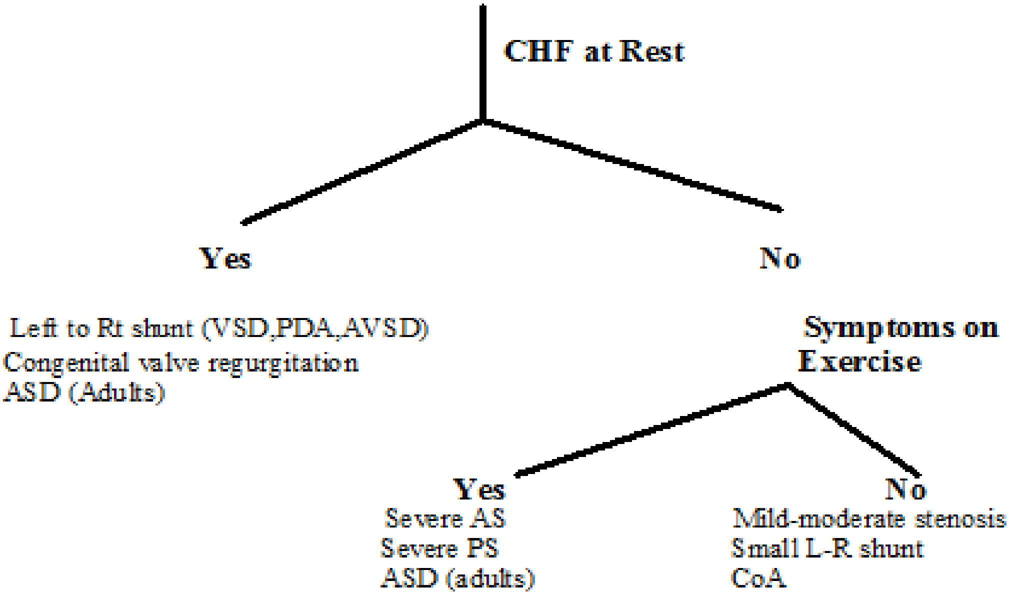
3-6 weeks	VSD, AVSD,PDA, TGA/VSD
1-2 Years	Non obstructed TAPVD, Partial APVD
3-7 Years	Ebstein disease, partial AVSD .
30-40 Years	ASD secundum

Variable age:

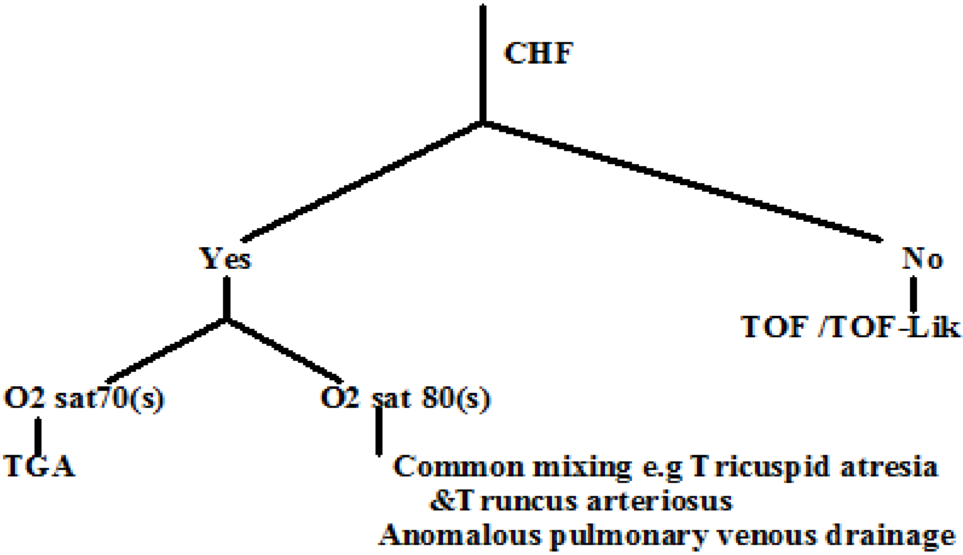
TOF, pulmonary and aortic stenosis depending on severity, partial AVSD,Ebstein anomaly of the tricuspid valve.

Classification of CHD by Inspection

1. Pink (O2 sat=>95%)



2. Cyanosed / Low O2 sat<95%


CHF
Yes
No
TOF /TOF-Lik
O2 sat70(s)
TGA
O2 sat 80(s)
Common mixing e.g Tricuspid atresia
&Truncus arteriosus
Anomalous pulmonary venous drainage

Common Case Scenarios

1. Infants with CHF and Normal O2 Saturation

History:

An eight months old infant presented with shortness of breathing, recurrent chest infections interrupted feeding and slow weight gain since the age of 2 months.

Physical examination:

A wasted infant with a weight of 5 kg. Distressed with respiratory rate of 50 breaths/min, heart rate 180 beats/min. O2 sat: 96%.

CVS:

Inspection:

Chest deformity: bilateral Harrison sulci, visible pulsations at the apex and the epigastrium

Palpation:

The apex beat is at the 6th intercostals space outside the mid clavicular line. Right ventricle (left parasternal) heave is positive, palpable second pulmonary sound at the second intercostals space. The liver is palpable 4 cm below the costal margin with a span of 8cm.Normal pulses were felt.

Auscultation: Normal S1, accentuated S2.An ESM at the left lower intercostals space (tricuspid area) associated with a soft mid diastolic apical murmur.

CXR: (Fig 7)

The CXR shows:

- Cardio thoracic ratio of 70%.
- LV and LA enlargement.
- Prominent main pulmonary artery.
- Prominent pulmonary arterial branches (pulmonary plethora).

ECG: (Fig 32)

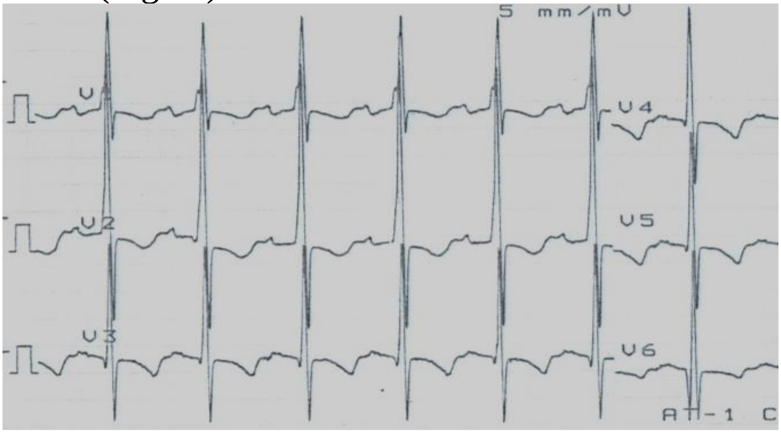


Figure 32: ECG of the patient showing biventricular and left atrial enlargement (Note the half standard mark), LV strain with inverted T wave in V6 is more typical of PDA than VSD.

Definitive Diagnosis:

Echo: showed a large mid muscular VSD 12 mm with Doppler gradient 10mmHg. LV diastolic dimension 3.4 cm (normal 2.4 cm) and left atrium to aorta ratio 2:1 (normal 1:1) (Figure 33)

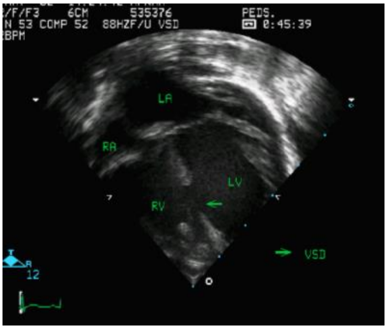


Figure 33 (above)
Echocardiographic 4 chamber view showing a large mid muscular VSD (arrow).

Management:

1. Diuretics: Start with furosemide then add spironolactone only if the signs of congestion (liver size) did not improve.No need for potassium supplement with this combination.
2. Angiotensine converting enzyme (ACE) inhibitors.
3. Add digoxin if heart failure is not controlled
4. High caloric formula: ready-made high caloric infant formula or concentrated regular infant formula. Advice frequent small feeds.
5. Surgery: the VSD is very large, at this age it is unlikely to close spontaneously.

Differential Diagnoses:

In infants with CHF and no cyanosis other possible diagnoses can be a large PDA, AVSD or cardiomyopathy. Auscultation, CXR and ECG can help to differentiate these conditions. (Figure 34,35 and 36) and echo is the definitive diagnostic test.

Hints to differentiate theses conditions:

1. A continuous (machinery) murmur at the infra-clavicular area indicates a large PDA. PDA murmurs can be long systolic with a short diastolic component. It can also be associated with a mitral mid diastolic murmur due to increased flow across the tricuspid valve as in VSD.
2. A short ESM at the tricuspid area with a pansystolic apical murmur indicates AVSD (associated with left AV valve regurgitation) AVSD is common in Down's syndrome

where it constitutes 50% of CHD in these patients. If the AVV is competent the physical signs will be similar to those with large VSD.

3. Pansystolic apical murmur with muffled heart sounds in cardiomyopathy (associated with functional MR)
4. **No murmur:**
   Can be found in dilated cardiomyopathy or a very large VSD. In the latter condition a loud P2, functional apical mid diastolic murmur and distinct heart sounds can help to differentiate from cardiomyopathy where the heart sounds are muffled.

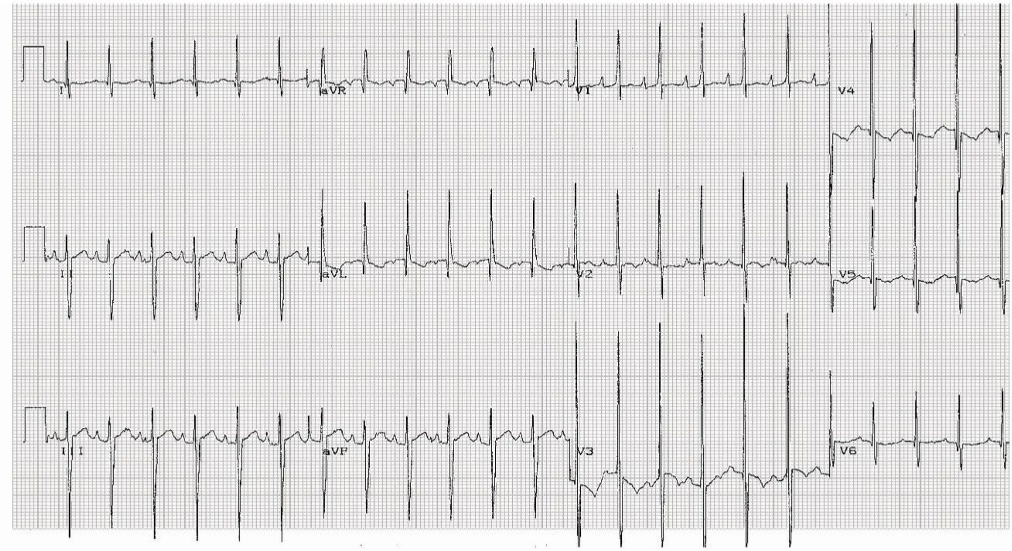


Figure 34

ECG of a patient with Down's syndrome and complete AVSD showing superior left axis (positive voltage in lead 1 and negative in AVF) and biventricular hypertrophy.

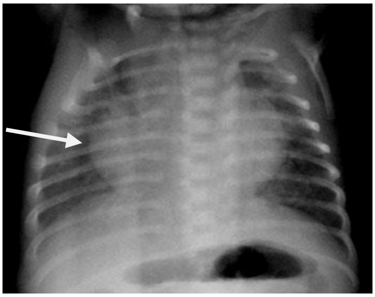


Figure 35
CXR from the same patient with AVSD showing cardiomegaly with right atrial(arrow) and ventricular enlargement with lung plethora. Compare with Figure 8.

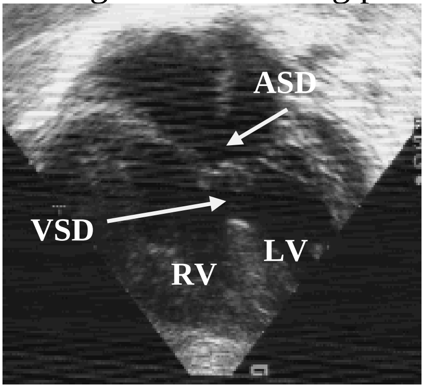


Figure 36
Echocardiographic 4-chamber view from a patient with complete balanced AVSD showing a large inlet VSD, large primum ASD and normal size LV and RV.

Discussion:
CHF is a clinical syndrome where the cardiac output is not sufficient to meet the body needs.

Chronological sequence of common causes of heart failure:

Age	Cause
In utero	Fetal arrhythmias
(hydrops fetalis)	Fetal cardiomyopathy
	Feta anemia (e.g. Rhesus incompatibility)
First week of life:	
Cyanotic	TGA, Truncus arteriosus
Acyanotic	Left side obstruction (CoA)
3-4 weeks	VSD, AVSD, PDA, SVT
Toddlers	TAPVD
Children	Rheumatic heart disease, MV prolapse
Adults	ASD
Any age	Myocarditis, cardiomyopathy

Pathophysiology of CHF:

Decreased cardiac output leads to activation of compensatory mechanisms that include sympathetic nervous system (adrenalin, noradrenalin) and neurohormonal activation (rennin-angiotensin-aldosterone) that leads to increased heart rate, vasoconstriction and fluid retention. These mechanisms lead acutely to increased cardiac output, however, in the long term, chronic sympathetic stimulation leads to cardiac toxicity, remodelling and worsening of CHF.

Clinical Features:

- Infants have characteristic triad of interrupted feeding, failure to thrive and respiratory distress. Recurrent chest infections are related to pulmonary congestion as well as compression of the bronchial tree by dilated PA branches and heart chambers.
- Older children present with effort intolerance.

Diagnosis:

- CHF is a clinical diagnosis, signs in infants are distinct from older children and include small weight, respiratory distress, enlarged heart chambers and hepatomegaly. Raised JVP, basal crepitations and lower limb edema are rare.
- CXR: Confirm the clinical diagnosis of cardiomegaly.
- Atrial natriureteric peptide levels (types A and B) had been documented to be a useful laboratory tests to diagnose CHF in children with different pathologies including CHD as well as cardiomyopathies. This test is particularly important in infants where respiratory distress can be due to pneumonia.

Management of CHF in infants:

- The first line drugs are diuretics which should be increased gradually adding potassium sparing drugs till the signs of congestion improve. When two diuretics are used care to electrolyte imbalance should be given. Potassium supplement are not routinely used unless there is documented hypokalemia.
- ACE inhibitors (enalapril and catptopril) should be used unless there is a contraindication like severe left or right ventricle outflow obstruction. AR and PDA are relative contraindications because of associated hypotension.
- Digoxin is added if there no improvement with the above drugs.
- Beta blockers (carvedolol) has proven to be effective in adults as well as children with CHF due to cardiomyopathy.

The time for surgical/ catheter intervention:

Depends on many factors:

1. For the most common types of VSD (perimembranous outlet and muscular trabecular types) there is a chance of spontaneous closure, however towards the second half of the first year (6-7months) the chances for spontaneous closure of large defects become low and surgery should be considered.

2. Certain types of VSDs like large inlet (same as in AVSD), malaligned and subarterial (doubly committed) types **do not close** spontaneously: so surgery is considered early if such defects are large.

3. Small VSDs should be followed up regularly: in the first year of life 4-6 monthly and later in life annually. The natural history of small VSD is generally favourable, however certain complications can occur:

i. AR can develop secondary to prolapse of the aortic valve cusp into the VSD leading to deformity of the vavle. The appearance of an early diastolic murmur should alert the physician to this complication, however mild AR should be looked for by echo and when found e surgical repair should be performed.

ii. RV outflow tract obstruction due to growth of a muscle bundle: clinically a new murmur (ESM at pulmonary area) may alert the physician. ECG changes from normal (as for small VSD) to RVH. Echo is diagnostic and the Doppler gradient will determine the need for surgery.

iii.Sub aortic membrane: careful echo examination with measurement of sub aortic gradient should be done.

iv.Infective endocarditis (IE): VSD is common cause of IE where vegitations commonly form on the TV leading to pulmonary septic emboli.

4. AVSD needs surgery: In complete AVSD there is a large VSD component and early presentation with CHF therfore surgery should be considered around 4-6 months of age. The incidence of Eisenmenger’s syndrome is early especially in patiets with

Down’s syndrome. In partial AVSD the VSD is tsmall/absent and timing of surgery is determined by the symptoms. If the valves are competent symptoms can be delayed to adolecnce and young adulthood while valve regurgitation leads to CHF earlier.Rarely, AVSD can exit with intact atrial and ventricular septa and the only manifestation is AV valve regurgitation.
5. Large PDA: most PDAs are closed by transcatheter device (page 46) except in young infants with very large PDAs. Device closure has a high rate of success in the short and long terms. Very small PDA without a murmur (silent PDA) is usually left alone.

Endocarditis prophylaxis is indicated for *all* left to right shunts except ASD. It is also indicated up to 6 months after successful closure of shunts.

Can CHD be prevented?
General measures: Avoidance of consanguineous marriage, strict control of diabetes before pregnancy and avoidance of viral infections e.g.by rubella vaccination can all lead to potential reduction in the incidence of CHD. The use of folic acid to prevent neural tube defects helped to test the efficacy of this drug in prevention of CHD. Many reports confirmed that the use of preconception folic acid supplementation by the mother significantly reduce the incidence of congenital heart defects in offspring.

Suggested Readings:

1.Soufflet V, Van de Bruaene A, Troost E, Gewillig M, Moons P, Post MC, Budts W. Behavior of unrepaired perimembranous ventricular septal defect in young adults. Am J Cardiol. 2010;105:404-7.

2.Gabriel HM, Heger M, Innerhofer P, Zehetgruber M et al. Long-term outcome of patients with ventricular septal defect considered not to require surgical closure during childhood. . J Am Coll Cardiol. 2002;39:1066-71.

3.Layangool T, Kirawittaya T, Sangtawesin C, Kojaranjit V et al. Natural aortic valve complications of ventricular septal defect: a prospective cohort study. J Med Assoc Thai. 2008;91 Suppl 3:S53-9.

4.Auerbach SR, Richmond ME, Lamour JM, Blume ED, Addonizio LJ, Shaddy RE, Mahony L, Pahl E, Hsu DT.BNP Levels Predict Outcome in Pediatric Heart Failure Patients: Post-hoc Analysis of the Pediatric Carvedolol Trial. Circ Heart Fail. 2010 Jun 23. (Pubmed)

5.Sulafa KM Ali. Cardiac abnormalities in children with Down's syndrome and their short term outcome. Cardiovas J South Afr 2009;20:112-115.

6.Sulafa KM Ali. Clinical and Echocardiographic features of Sudanese patients with atrioventricular septal defects and their short term outcome. Khartoum Medical Journal 2009;2:165-168.

7. Cantinotti M, Vittorini S, Storti S, Prontera C et al. Diagnostic accuracy and clinical relevance of brain natriuretic peptide assay in pediatric patients with congenital heart diseases. JCardiovasc Med (Hagerstown). 2009;10706-13.

8. van Beynum IM, Kapusta L, Bakker MK, den Heijer M, Blom HJ, de Walle HE. Protective effect of periconceptional folic acid supplements on the risk of congenital heart defects: a registry-based case-control study in the northern Netherlands. Eur Heart J. 2010 Feb;31:464-71.

2. Low O2 Saturation *without* CHF:

Case History:

Scenario A

A 12 months old boy gave history of episodes of shortness of breathing labelled as 'chest infections' with recurrent admissions for antibiotics and O2.

Physical Examination

The patient is well with mild duskiness, O2 sat of 85%.There is clubbing. No respiratory distress. The weight is 10 kg.
CVS: a harsh ESM 3/6 was heard at the pulmonary area, no signs of CHF.

Scenario B

The same patient as in A presented today to the emergency room with low grade fever, shortness of breathing and increasing cyanosis.

Physical Examination:

In the emergency room the patient was found to be ill, limp, in respiratory distresse, Respiratory rate was 50/minute and heart rate 150/minute. O2 sat 70% on 5 L/min O2. No murmur heard on auscultation.

CXR (Fig6)

1. Small heart
2. Oligemic lungs

3. Small main pulmonary artery with rounded raised apex (boot shaped)

ECG: (Fig 37)
Right ventricle hypertrophy

Echo: TOF. Large malaligned VSD, overriding aorta by 50%, severe pulmonary vavle and subvalvre stenosis with a gradient of 70mmhg.(Figure 38)

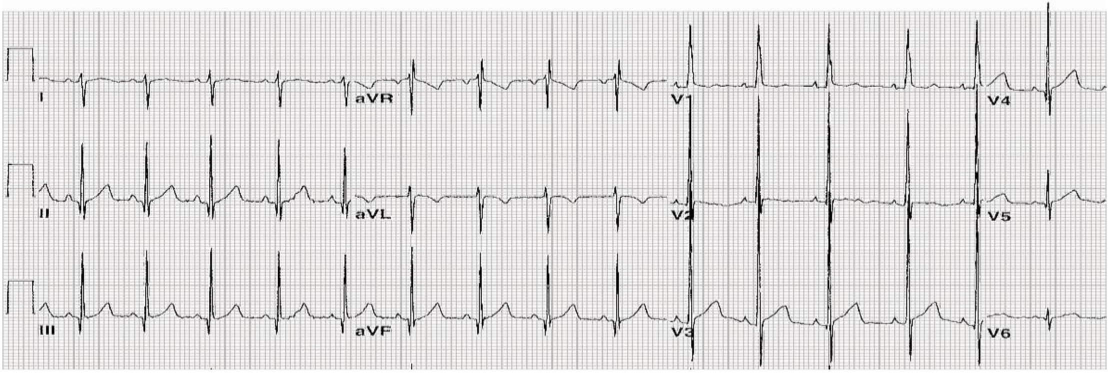


Figure 37:
ECG of a patient with TOF showing RVH (pressure overload with tall R and upright T wave in V1)

Echo: TOF. Large malaligned VSD, overriding aorta by 50%, severe pulmonary vavle and subvalvre stenosis with a gradient of 90mmhg.(Figure 38)

Diagnosis:
Cyanotic Spell due to TOF

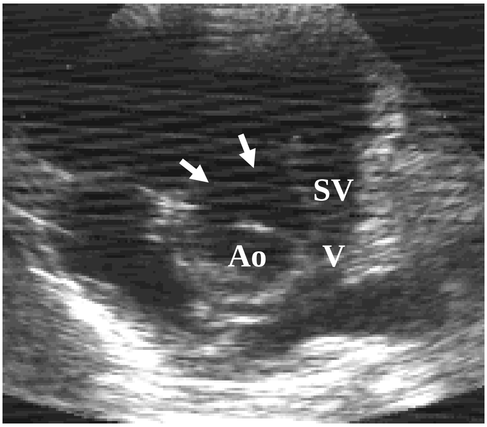


Figure 38:
Echocardiographic short axis view showing the aorta (Ao), a large malaligned VSD (arrows) and a narrow RV outflow tract with valve (V) and sub-valve (SV) stenosis.

Management:

Scenario A

1. **Prophylactic oral propranolol:** 0.2- 0.4 mg/kg/dose TDS, increased gradually to 1.0-1.5 mg/kg/dose TDS (watch for hypotension and hypoglycaemia**).**
2. Keep hemoglobin =>12gm/dl with normal indices.
3. Surgical correction.

Scenario B

Management:

1. The patient should be kept comfortable on the mother's lap in his/her preferred position.

2. The knee-chest position is helpful but *don't force the child*.
3. Oxygen
4. Subcutaneous morphine before attempting IV line (dose 0.1mg/kg/dose)
5. IV line is put in and blood is sent for complete blood count.
6. **IV fluids-** isotonic saline or colloid bolus of 10-20cc/kg by rapid IV push which may be increased to 60cc/kg. Extra volume can be given in cyanotic spell as the physiology does not induce CHF.Continue full maintenance IV fluid.
7. Morphine can be given as regular dose 0.05mg/kg/dose IV q 8hours
8. Blood gas: treat metabolic acidosis, can treat with bicarbonate empirically (dose 1meq/kg)
9. Treat the precipitating factor: fever, dehydration, infection
10. Keep Hb => 12 g/dl.
11. Propranolol: oral propranolol should be started. IV propranolol can be considered but serious side effects like bradycardia and hypotention should be anticipated.

Further options:

1. **Metoprolol** is given under BP and ECG monitoring at a loading dose of 0.1mg/kg intravenously over 5 minutes, repeat every 5 min to max 3 doses , then start infusion 1-5 mcg/kg/min. Metoprolol is shorter acting than propranolol hence in case of significant bradycardia stopping the infusion will reverse the effect in 30 minutes to 1 hour.
2. **Esmolol** 0.5 mg/kg over 1 minute then 0.05 mg/kg/min for 4 minutes. This is an ultra-short acting cardioselective beta blocker, should be used

cautiously via peripheral IV line as extravasations can cause severe tissue necrosis.

3. **Intravenous Phenylephrine:** This is an alpha adrenergic blocker which increases the systemic vascular resistance. Dose**:** 0.01 mg/kg IV slowly or 0.1mg/kg subcutaneously. The dose should be titrated to the BP response.
4. **Ketamine:** 0.25-1.0 mg IV or IM, has dual benefit causes sedation and increase systemic vascular resistance.
5. General anesthesia and emergency surgical intervention (palliation/repair).

- Prophylactic oral propranolol should be used in all patients with cyanotic spells till surgery is done**.**

Discussion:

- Cyanotic spell is the most common presenting feature of TOF and TOF like conditions to emergency room.
  It is often misdiagnosed as 'chest infection' or more seriously 'heart failure' (see table 11). It is mandatory to differentiate the two as diuretics can have deleterious effects in cyanotic spells leading to increasing blood viscosity and thrombosis.
- TOF is not associated with heart failure. If heart failure is suspected another additional or alternative diagnosis should be carefully sought like associated TAPVD or severe anemia.
- A common error is to diagnose patients with a large malaligned VSD plus overriding of aorta associated with mild or no PS as TOF. This type of anatomy is considered the same in pathophysiology as a large VSD as mild PS does not protect the lungs therefore the patient will present as CHF rather than cyanotic spell.

- Many conditions have a TOF-like pathophysiology: Pulmonary atresia/VSD,tricuspid atresia with PS, TGA/VSD with PS, AVSD with PS can all present with cyanotic episodes similar to TOF especially when associated with infundibular stenosis. The medical treatment is generally similar to TOF.
- **Pulmonary atresia/VSD** is the extreme type of TOF and present earlier. Clinically it can be suspected by the absence of PS murmur. The source of pulmonary blood flow is determined by echo, CT/MRI or cardiac cath. Echo can show multiple aorto-pulmonary collaterals which can rarely lead, if large enough, to pulmonary overflow manifesting clinically as heart failure. Angiography is usually needed to delineate the anatomy of PA branches and suitability for surgical correction. When the central PA branches are hypoplastic and collaterals widely separated surgery can be challenging.
- The pulmonary stenosis murmur often disappears during the spell because of severe sub pulmonary stenosis (infundibular spasm) and comes back after treatment.
- **Pink TOF:** This type of TOF is characterized by normal or slightly reduced O2 sat and no clubbing. The anatomy of pink TOF patients is the *same* as those with cyanosis i.e the degree of pulmonary stenosis is severe however, the degree of cyanosis is mild probably due to a balance between systemic and pulmonary vascular resistance favouring adequate pulmonary blood flow. Cyanotic spells *do* occur in pink TOF when this balance is disturbed or when the infundibular stenosis increases.
- The actual cause of cyanotic spell is uncertain. Possible aetiologies include an imbalance between systemic and pulmonary vascular resistance leading to decreased pulmonary flow. Factors that decrease systemic resistance

i.e. cause hypotension include fever, dehydration, pain and crying and all can precipitate spells.

- Treatment of spell (detailed above) aim to increase the systemic resistance (by fluids, ketamine, phenylephirine) and decrease the pulmonary vascular resistance (by O2).
- **TOF with absent pulmonary valve** is one of the unique types of TOF. It is characterized by a dysplastic PV with significant regurgitation leading to severe dilatation of main pulmonary artery and its branches (Figure 39). The clinical features are variable: patients can present in infancy with respiratory symptoms due to compression of the airway by the dilated pulmonary arteries (Figure 10). Physical examination can identify this syndrome by careful auscultation for a ‘to and fro’ murmur indicating pulmonary stenosis and regurgitation. Surgical repair is done by VSD closure and a pulmonary – right ventricle valved conduit.
- **TOF with AVSD**: is a known association especially in Down’s syndrome. The ECG is characteristic (Figure 41)
- Surgical management of TOF: depends on the onset of symptoms and the availability of open heart surgery. If symptoms occurred early and open heart surgery is not possible palliation by modified Blalock-Taussig shunt should be done. Refinement of surgical techniques includes the preservation of pulmonary valve annulus (avoiding trans-annular patch) in order to decrease post operative pulmonary regurgitation (see scenario 15-B) and avoidance of ventriculotomy. In well equipped centres surgical repair can be done as early as the neonatal period.
- Lifelong follow up and endocarditis prophylaxis are needed in patients who underwent TOF repair.

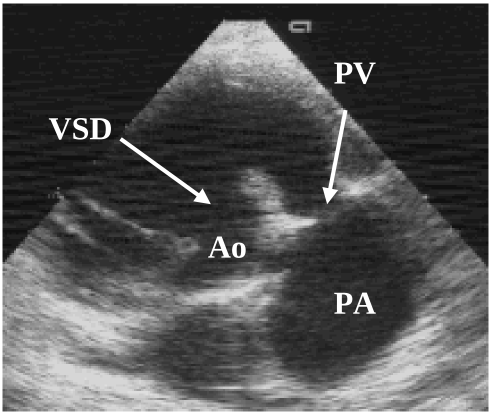


Figure 40:
Echocardiographic short axis view showing a dysplastic pulmonary valve (PV) and dilated pulmonary artery (PA) in a patient with TOF-Absent pulmonary valve syndrome.

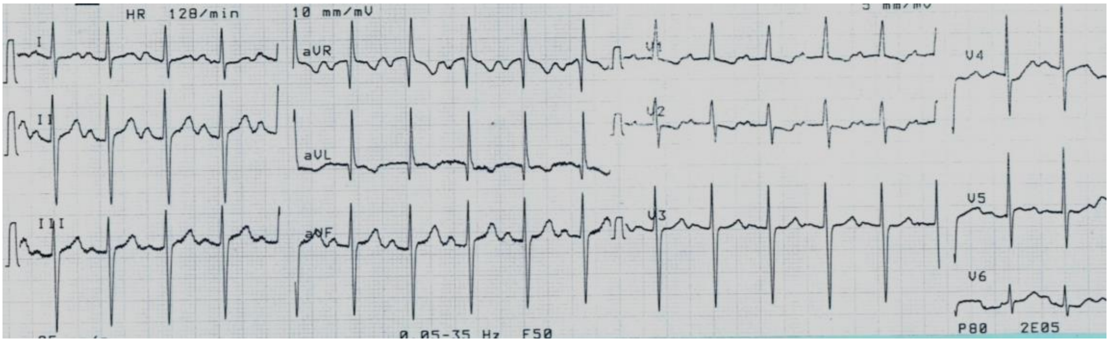


Figure 41:
ECG of a patient with Down's syndrome with clinical cyanosis and clubbing. There is superior left axis with RVH. Similar findings can be present in AVSD complicated by Eisenmenger's syndrome.

Table 11:

Comparison between Cyanotic Spell and Heart failure

Cyanotic Spell (TOF /TOF like)	**Cyanosis with heart failur**e (e.g.TGA)
Tachypnea	Tachypnea
Normal weight	Wasting and failing to thrive
No chest deformity	Can have chest deformity
Apex not displaced	Apex can be displaced
Liver not palpable	Liver is palpable
Chest XR: Small heart, oligemic lungs	Large heart with plethoric lungs

Suggested Readings:

1.Zucker N, Rozin I, Levitas A, Zalzstein E. Clinical presentation, natural history, and outcome of patients with the absent pulmonary valve syndrome. Cardiol Young. 2004 ;(4):402-8
2.Sulafa KM Ali. Unusual presentations and anatomy of Tetralogy of Fallot in Sudanese patients. Khartoum Medical Journal 2008;1:125-28

2.Kaza AK, Lim HG, Dibardino DJ, Bautista-Hernandez V et al.Long-term results of right ventricular outflow tract reconstruction in neonatal cardiac surgery: options and outcomes. J Thorac Cardiovasc Surg. 2009;138:911-6.

3.Heart Failure *and* Low O2 Saturation

Scenario A:
Cyanotic CHD presenting in early infancy:
A 2 month old baby presented with respiratory distress, blue discoloration of lips and interrupted feeding since birth which is increasing gradually.
P/E
The baby is cyanosed; O2 saturation is 70% in room air and 80% on O2. Wt is 3.5 kg. Respiratory rate is 65 breath/ minute and heart rates 170 beats per minute. Normal S1 and single S2 are heard. A short systolic murmur is heard at the left lower sternal border. The liver is enlarged 5 cm below the costal margin with a span of 8 cm.

CXR:
Cardiomegaly with' egg on side' appearance (Figure 12)
Narrow heart base
Plethoric lungs

ECG: Normal
Normal right axis for age
RVH (appropriate for age)
Echo : The aorta is anterior and to the right of pulmonary artery (D-TGA). A perimembraneous VSD 3 mm with Doppler gradient 40mmhg. Normal other anatomy.
Diagnosis: TGA with VSD.

Management:

1. Oxygen.
2. Diuretics.

3. Referral for surgical repair.

Differential Diagnosis:
Cyanotic CHD with heart failure in infants:
This group includes 2 types of cyanosis:

1. **Severe cyanosis with CHF**: O2 saturation in the range of 60-70%: usually due to simple TGA or other complex lesion with TGA and *no* PS (e.g Double outlet RV with sub pulmonary VSD.
2. **Milder cyanosis** with CHF: In this category of patients the O2 saturation is 85-90% and cyanosis can be missed clinically. Differential diagnoses include truncus arteriosus (Figure 42), all lesions with functionally single ventricle *without* PS (e.g. tricuspid atresia, double inlet LV) and total anomalous pulmonary venous drainage. Tricuspid atresia can be identified from the ECG.(Figure 83)

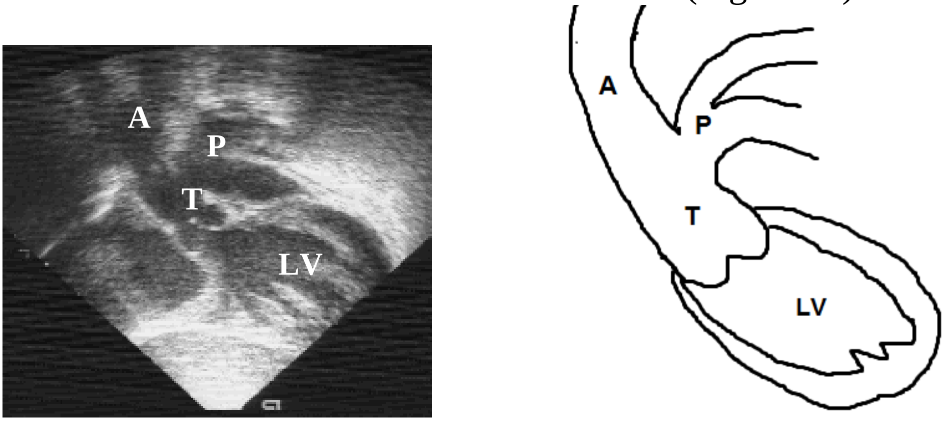


Figure 42:
Left: Echocardiographic subcostal view showing a single arterial trunk (T) branching into the pulmonary artery (P) which has 2 branches and the aorta (A) which is arching. Right: schematic presentation for the echo image.

Non-cardiac Causes:

Severe lung disease can cause a similar clinical scenario. The response to oxygen (hyperoxia test) can be helpful.

Hyperoxia Test:

The arterial blood gas is done before and after giving100% O2 for 10 minutes. If PO2 increases to > 200 mmHg, cyanotic CHD are ruled out. In patients with common mixing conditions (truncus arteriosus and functionally single ventricle) the PO2 may increase modestly up to 100 mmhg. If PO2 remains below 70mmhg severe cyanotic CHD (like TGA and pulmonary atresia) or very severe lung disease could be the cause.

Pulse oximetry can be helpful as a non invasive tool for the hyperoxia test instead of doing arterial blood gas, however the blood gas has the advantage of reading the PCO2 which is high in pulmonary disease.

Differential diagnosis of a PO2 < 70mmhg on 100% O2:

1. TGA, Pulmonary atresia, severe TOF
2. Severe respiratory disease e.g. acute (adult) respiratory distress syndrome (ARDS), congenital lung anomaly or hypoplastic lungs.
3. Severe pulmonary hypertension.

Chest Xray can help to differentiate pulmonary from cardiac causes.

Definitive Diagnosis: Echo.

Discussion:

Early diagnosis is crucial in CHD presenting with cyanosis and CHF early in infancy. Antenatal (fetal echo) and early neonatal examination can help to diagnose such lesions so patients can be referred for early surgical repair/palliation. In TGA/VSD and truncus arteriosus early surgical repair in the first 2 months of age should be performed. In tricuspid atresia and other conditions with functionally single ventricle early palliation by pulmonary artery banding is indicated. As such patients do not have pulmonary stenosis their pulmonary vascular bed is not protected and they develop pulmonary vascular obstructive disease (Eisenmenger's syndrome) in the first few months of life.

Cyanotic CHD presenting later in infants/toddlers

Scenario B:

A 2 year old boy presented with shortness of breathing for 6 months which is increasing gradually.

P/E

Weight 8 kg, no clear cyanosis, mild clubbing.
CVS: RV heave, palpable P2, short ESM at pulmonary area.
Liver 4cm below costal margin with a span of 9 cm.O2 sat 85%.

Chest XR

Cardiomegaly, RVH, plethoric lungs

ECG (Fig 43)

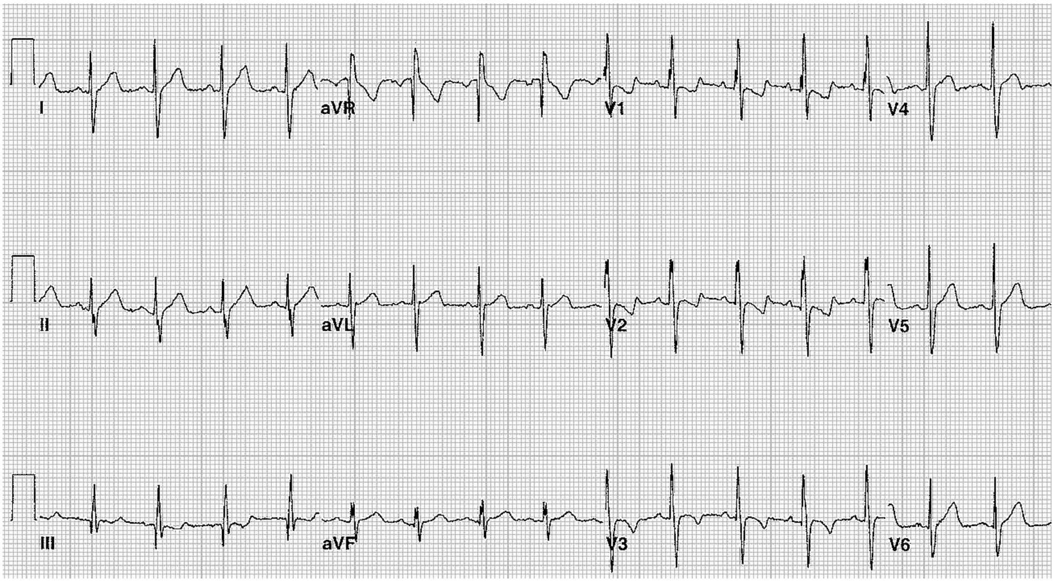


Figure 43:
ECG showing RVH volume overload (Rsr) and right axis deviation
Differential diagnoses: ASD secundum.

Echo:
Total anomalous pulmonary venous drainage to the coronary sinus (cardiac type). The 4 pulmonary veins were seen draining to an enlarged coronary sinus.The RV is dilated. Small ASD secundum with right to left shunt.
(Figure 44)

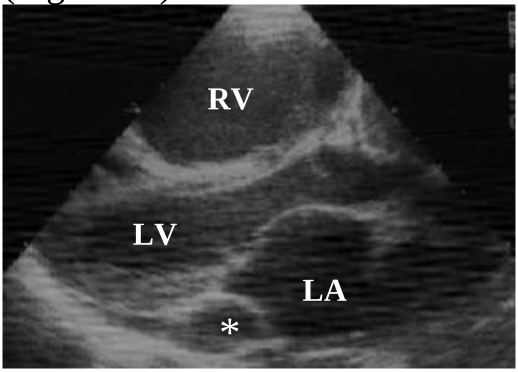


Figure 44 (above):
Parasternal long axis view showing dilated RV and coronary sinus (astrix). This should alert the echocardiographer to the diagnosis of TAPVD.

Management:
Diuretics
Surgical correction.

Scenario C:
An 8 year old boy presented with SOB on severe exertion for 6 month.
P/E
Cyanosed, clubbed. O2 saturation 80%. Stunted growth. Increased jugular venous pressure. Auscultation: S1, split S2 and S3 present (triple rhythm). PSM at left lower sternal border. The liver is 6 cm below the costal margin (span 10 cm).
CXR: Figure 45

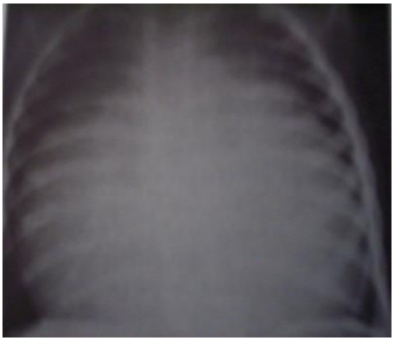


Figure 45
CXR showing massive cardiomegaly with right atrial enlargement

Differential diagnosis of massive cardiomegaly:

- Pericardial effusion
- Ebstein’s anomaly
- Cardiomyopathy
- Multiple valvular lesions
- Long standing severe pulmonary valve stenosis

ECG: Fig 46

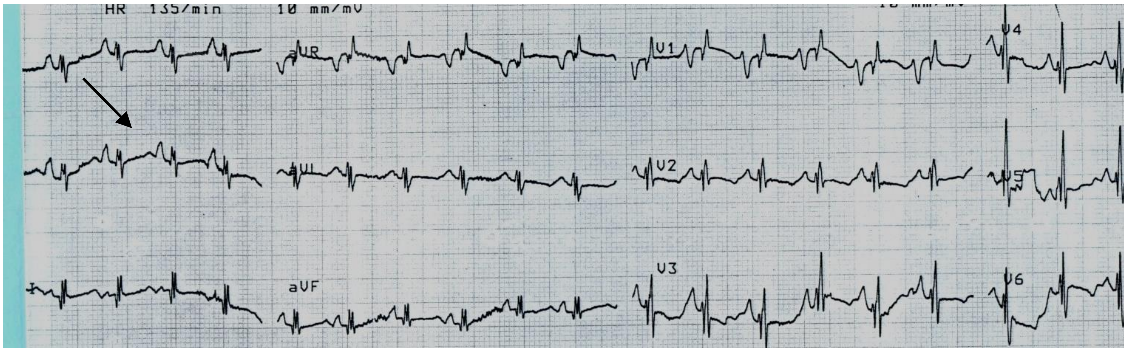


Figure 46:
ECG showing a tall P wave (arrow), right axis deviation and rsR pattern in a patient with Ebstein’s anomaly.Possible qR pattern in V1 indicating RV volume overload.

Echo: Ebstein anomaly of the TV (Figure 47). Displacement of the septal leaflet of the TV to the RV apex by 40mm/m2. Sever TR with normal velocity (normal pulmonary artery pressure), ASD secundum 8 mm with right to left shunt.

Management:

1. Frusemide , spironolactone
2. Discussion with the surgeon.

Discussion:

Cyanosis and heart failure in toddlers and children can be due to:

1. **Anomalous PVD:**

In TAPVD the O2 saturation is 85-90% and cyanosis can be missed. Supracardiac APVD is the most common type and can be identified from CXR if a'figure of 8' appearance is present, usually in older children (Fig13).

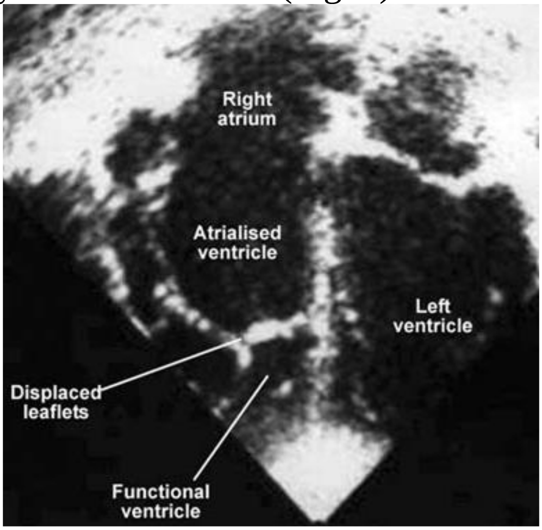


Figure 47:

Echocardiographic 4 chamber view from a patient with Ebstein's anomaly showing markedly displaced TV leaflets.

The anomalous veins join the innominate vein and superior vena cava through an ascending (vertical) vein. The 'figure of 8' appearance is due to the dilated vertical vein on the left and the superior vena cava on the right supracardiac borders. Other types of APVD are the cardiac type where the veins drain to the coronary sinus as in scenario B, to the superior vena cava or directly to RA. The coronary sinus is best seen in the subcostal, parasternal long and 4 chamber views. The infracardiac type (the least common) presents early in the neonate as it is commonly associated with obstruction. It is considered a surgical emergency with a high surgical mortality .TAPVD is almost always associated with an ASD. Partial APVD causes milder symptoms similar to ASD so patients preset in the 2nd or 3rd decade of life. A specific type of ASD (sinus venosus ASD) which is a defect in the most superior part of the atrial septum is associated with anomalous right upper pulmonary vein to superior vena cava. Secundum ASD can be associated with partial APVD therefore pulmonary veins should be carefully evaluated by echo in patients with ASD. Scimitar syndrome constitutes lung sequestration (usually the right lower lobe) which is supplied by an aortic collateral, the sequestered lobe has an anomalous PVD to the inferior vena cava. It has a characteristic CXR appearance (figure9). Echo and CT/MRI confirm the diagnosis. The main treatment is ligation or catheter closure of the aortic collateral and in selected cases resection of the sequestered lobe.

2.**Ebstein's anomaly of TV:**

Is defined by apical displacement of the septal leaflet of the tricuspid valve by > 8mm/m2, dilated right atrium with atrialization and variable degrees of hypolasia of the RV. It is commonly associated with ASD, the right to left shunt across the ASD is due to the TR rather than increased PA pressure, i.e. the PA pressure is normal. It has a wide spectrum of clinical presentations: the most severe form presents in the neonatal period with CHF and cyanosis while some patients remain asymptomatic for long periods and present in adulthood with mild TR,cyanosis and/or CHF. Many patients present with SVT due to accessory pathway that can be detected on their ECG as WPW syndrome (see page117) or Mahaim-type preexcitation (Figure 48). The frequency of arrhythmia in paediatric patients with Ebstein's disease is reported as 17% and increases as the patient grows into adulthood. The frequency of Ebstein's disease in Sudan (2.5% of CHD) is much higher than the frequency reported in the Western literature which is 0.5%. Management is challenging and needs to be individualized according to the age and symptoms of the patient and the cardiac anatomy. Valve repair needs extensive expertise and the results of valve replacement are not encouraging. Occasionally a large ASD with dominant left to right shunt can be closed by device, however it is better to investigate for accessory pathways (invasive electrophysiological studies) prior to catheter closure. Some patients with severely displaced valves may benefit from Glenn shunt palliation.

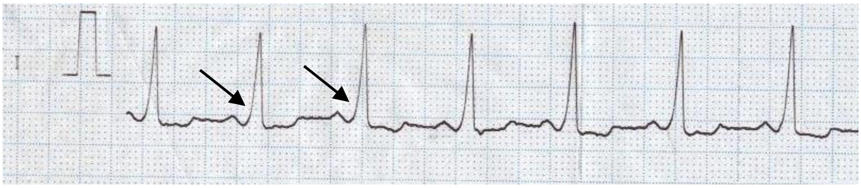


Figure 48:
Rhythm strip from a patient with Ebstein anomaly and Mahaim type preexcitation. Note the delta wave (arrows) and relatively normal PR interval.

3. **Pulmonary hypertension (PHT):**
   Is defined as pulmonary artery mean pressure > 25 mmhg and pulmonary vascular resistance (PVR) >3 Wood units. The global prevalence of PHT is hard to estimate because accurate diagnosis of PHT is difficult, and access to care is limited in many countries. Because diseases that are risk factors for PHT, such as HIV, schistosomiasis, and sickle cell disease, are more prevalent in the developing world, the global burden of PHT is likely greater than is

recognized currently It has a wide range of aetiological factors.

WHO classification of PHT:

1. Pulmonary arterial hypertension (PAH)
1.1. Idiopathic PAH
1.2. Heritable
    1.2.1. Bone morphogenetic protein receptor type
    1.2.2. Activin receptor-like kinase type 1$_{;}$, endoglin (with or without hereditary hemorrhagic telangiectasia)
    1.2.3. Unknown
1.3. Drug- and toxin-induced
1.4. Associated with
    1.4.1. Connective tissue diseases
    1.4.2. HIV infection
    1.4.3. Portal hypertension
    1.4.4. Congenital heart diseases
    1.4.5. Schistosomiasis
    1.4.6. Chronic hemolytic anemia
1.5 Persistent pulmonary hypertension of the newborn
1. 6 Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)

2. Pulmonary hypertension owing to left heart disease
2.1. Systolic dysfunction
2.2. Diastolic dysfunction
2.3. Valvular disease
3. Pulmonary hypertension owing to lung diseases and/or hypoxia
3.1. Chronic obstructive pulmonary disease
3.2. Interstitial lung disease

3.3. Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4. Sleep-disordered breathing
3.5. Alveolar hypoventilation disorders
3.6. Chronic exposure to high altitude
3.7. Developmental abnormalities.

4.**Chronic thromboembolic pulmonary hypertension (CTEPH)**

5.Pulmonary hypertension with unclear multifactorial mechanisms
5.1. Hematologic disorders: myeloproliferative disorders, splenectomy
5.2. Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis: lymphangioleiomyomatosis, neurofibromatosis, vasculitis.
5.3. Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders.
5.4. Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis

Pathophysiology of Primary Pulmonary HTN:

Endothelins are a family of three peptides of 21 amino acids with strong vasoconstrictor effects. The three peptides are encoded by three different genes and derived from precursors (" big endothelins") which are cleaved by metalloproteases, named endothelin-converting enzyme. The endothelin system participates in the development of atherosclerosis as well as pulmonary hypertension, and mediates cardiac remodeling in heart failure. Five concepts provide a framework to understand and treat PHT:

(1) Endothelial dysfunction creates an imbalance that favours vasoconstriction, thrombosis, and mitogenesis. Most current therapy aim at restoration of this balance by inhibition of endothelin and thromboxane or augmentation of nitric oxide (NO) and prostacyclin.

(2) PHT has a genetic component. Mutations (bone morphogenetic protein receptor-2 [BMPR2]) and single-nucleotide polymorphisms (SNPs; ion channels and transporter genes) predispose to PAH.

(3) Excess proliferation, impaired apoptosis, and glycolytic metabolism in pulmonary artery smooth muscle, fibroblasts, and endothelial cells suggest analogies to cancer. Many experimental therapies reduce PAH by decreasing the proliferation/apoptosis ratio; these include inhibitors of pyruvate dehydrogenase kinase (PDK), serotonin transporters (SERT), survivin, 3-hydroxy-3-methylglutaryl coenzyme A reductase, transcription factors and nuclear factor of activated T lymphocytes. Tyrosine kinase inhibitors used to treat cancer are currently in phase 1 PAH trials.

(4) Refractory vasoconstriction: While fewer than 20% of PAH patients respond to conventional vasodilators, refractory vasoconstriction may respond to kinase inhibitors.

(5) The RV can be targeted therapeutically by using diuretics.

Diagnosis:

- Clinical diagnosis: Effort intolerance, syncope or edema can be the presenting symptoms. Examination reveals subtle signs in early cases, right sided heart failure (increased jugular venous pressure, hepatomegaly and lower limb edema), and variable degrees of desaturation, loud P2 and pan systolic murmur of TR can be present.
- ECG is a sensitive tool for PHTN showing RVH and right atrial enlargement.
- Echo: reliable in quantitative assessment of PHTN. TR peak systolic gradient accurately estimates the PA peak

systolic pressure and pulmonary regurgitation can estimate the PA end diastolic pressure.RV enlargement, abnormal position of the interventricular septum and right to left shunting through foramen ovale can be present.

- Cath: the gold standard investigation, PA pressure and pulmonary vascular resistance are directly measured, and the anatomy verified.Reversibility testing using O2 and nitric oxide are done.
- Investigations to find a cause of PHTN are needed before labelling the patient as primary PHT.

Management:

Most current treatments (prostacyclin, endothelin antagonists, and warfarin) address endothelial dysfunction by augmenting vasodilator and antiproliferative mediators and inhibiting vasoconstrictor, prothrombotic, and mitogenic pathways. Treatment of PAH involves the use of prostanoids (given intravenously, by inhalation, subcutaneously, or orally), endothelin receptor blockers (Bosentan), and/or phosphodiesterase 5 inhibitors (sildenafil). The prognosis is guarded (see scenario 5).

Suggested Readings:

1.Basso ML, Pacheco GC, Netto OS, Abuchaim JS et al.Total anomalous pulmonary venous drainage: 29 years of experience in a single institution. Arq Bras Cardiol. 2010;94:608-12.

2.Sulafa K.M.Ali, Nuha A.G.M.Nemiri. Echocardiographic and clinical features of Ebstein malformation in Sudanese children. Cardiol Young 2006; 16: 147–151

3.Delhaas T, Sarvaas GJ, Rijlaarsdam ME, Strengers JL et al.Long-term study on arrhythmias in children with Ebstein anomaly. Pediatr Cardiol. 2010;31:229-33.

4.Chauvaud S. Ebstein's malformation. Surgical treatment and results. Thorac Cardiovasc Surg 2000; 48: 220–223.

5.Simonneau G, Robbins IM, Beghetti M, Channick RN, Delcroix M, Denton CP, Elliott CG, Gaine SP, Gladwin MT, Jing ZC, Krowka MJ, Langleben D, Nakanishi N, Souza R. Updated clinical classification of pulmonary hypertension. *J Am Coll Cardiol.* 2009; 54: S43–S54.

6.Stephen L. Archer, E. Kenneth Weir,Martin R. Wilkins. Basic Science of Pulmonary Arterial Hypertension for CliniciansNew Concepts and Experimental Therapies. Circulation. 2010;121:2045-2066.

4. The Sick Neonate

The sick neonate is a very special situation where neonatology, cardiology and general paediatrics meet and coordinate.

The Fetal Circulation:

- The site for gas exchange is the placenta which has a low vascular resistance.
- The lungs are not inflated therefore they have a high vascular resistance.
- There are 4 physiological shunts in the fetus:

1. The placenta
2. The sinus venosus which diverts the oxygenated blood from the liver to the inferior vena cava.
3. The foramen ovale which shunts blood from the right to the left atrium.
4. The ductus arteriosus which diverts blood from the pulmonary artery to the aorta.

- The maximus partial pressure of oxygen in the fetal blood is 35 mmHg found in the umbilical vein.

The course of Fetal Circulation:

- The majority of bood from the umbilical vein , bypasses the liver through the ductus venosus to the inferior vena cava then to the right atrium.
- Blood mixes in the right atrium with venous return coming from the superior vena cava.
- The oxygenated blood is directed by the eostachian valve of the inferior vena cava to the foramen ovale to enter the left atrium, this portion represents 80% of the blood in the right atrium.

- Blood passes to the left ventricle then the aorta, where it mixes with blood from the ductus arteriosus in the desending aorta. Therefore the head and neck are supplied by more oxygenated blood than the lower parts.
- The remaining 20% of right atrium blood passes to the RV then PA, most of it is diverted by the ductus arteiorsus to the decending aorta.
- Blood returns to the placenta via the umbilical arteries.

The Transitional Circulation:

- With ligation of the umbilical core and the first breath the low resistance placenta is removed therefore the systemic BP increases and the pulmonary pressure falls, this leads to the following:

  1. Breathing and decrease in pulmonary resistance lead to increase pulmonary flow and increase of arterial oxgen concentration
  2. Increase in oxygen concentration leads to physiological then anatomical closure of the ductus arteriosus.
  3. Increase in left atrim pressure leads to physiological closure of the foramen ovale.
  4. Ligation of the umbilical vein leads to gradual closure of the ductus venosus.

Post natal Examination:

Thorough physical examination after birth and before discharge from hospital is very important.

- General examination to look for congenital defects in all systems is needed.

Evaluation of the Cardiovascular System:

Pulse oximetry:

-Pulse oximetry improves the detection of cyanotic CHD and should be considered as a routine screening test. It should be done in upper and lower limbs

- O2 sat is better be done after 24 hours of age to avoid false positive results which can be up to 5%.
- Upper limb (pre ductal) and lower limb (post ductal) saturation difference should be not more than 5%. If more then left side obstructive lesions have to be ruled out.
- Saturation above 95% is considered normal

Auscultation:

- Large left to right shunts can be murmur-less or with soft ejection systolic murmurs due to the high pulmonary resistance at this age.

Murmurs at the pulmonary area:

- Pulmonary stenosis murmur: a harsh long murmur indicates severe pulmonary stenosis.
- Soft murmurs: usually indicate physiologic pulmonary artery branch stenosis (innocent murmur) when it radiates to the back.
- If the murmur is moderately harsh consider significant PS which can be underestimated due to the high pulmonary resistance.

Murmurs at the left lower sternal board:

- VSD murmurs: if the VSD is large the murmur can be absent /soft
- Small VSD: murmur can be soft or harsh
- Tricuspid regurgitation: a harsh pansystolic murmur indicates tricuspid regurgitation which is usually secondary to pulmonary hypertension.

- Primary tricuspid valve disease: Ebstein’s anomaly can present at birth, the patient is usually cyanosed with heart failure and huge cardiomegaly.

PDA murmur:

PDA is common in the first few days of life. The murmur is usually short systolic with or without a diastolic component.

The Pulses:

- Palpate the brachial and femoral pulses and compare the volume.

Coarctation of the aorta is a neonatal emergency

- Absent/ week femoral pulse with normal brachial pulse indicates coarctation of the aorta.
- Normal femoral pulse can be present if there is coarctation with a large PDA.O2 sat difference of > 5% can be detected in this type of patient.
- Weeak left brachial pulse can occure if CoA involves the left subclavian artery.

Blood pressure:

Using suitable cuff measure the upper (right arm) and lower limb blood pressure. A difference of > 15 mmHg (higher in upper limbs) indicates CoA.

PDA- Dependant Circulation:

1.PDA- dependant pulmonary blood flow:

When the pulmonary blood flow is critically reduced e.g. in pulmonary atresia, severe TOF and criticl PS the PDA is essential to supply blood to the PA from the aorta (left to right shunt).

2.PDA dependant systemic blood flow:

In severe CoAo, HLHS and critical AS the blood flow to the systemic circulation depends on right to left (from PA to aorta) shunt across the PDA.

3**.PDA dependant mixing:**

In TGA the systemic and pulmonary circulations are parallel therefore PDA shunt is needed for mixing, in this situation the shunt is bidirectional.

- In the above situation the PDA needs to be open by using prostaglandin E1 intravenous infusion.

The starting dose is 0.05 to 0.1 μg/kg per minute, administered in a continuous intravenous drip. When the desired effects (increased PO2, increased systemic blood pressure, improved Ph) are achieved, the dose should be reduced step by step to 0.01 μg/kg per minute. When the initial starting dose has no effect, it may be increased up to 0.4 μg/kg per minute.

Three common side effects of intravenous infusion of PGE1 are apnea (12%), fever (14%), and flushing (10%). Less common side effects include tachycardia or bradycardia, hypotension, and cardiac arrest.

Scenario A
Collapsed Neonate with poor pulses:

A 3 day old infant was brought to ER with severe respiratory distress, drowsiness and dusky colour.

P/E

Week pulses all over, absent femoral pulse, HR 170, RR 80, O2 sat in upper limbs is 95% and in the leg 82%. Faint ESM is heard. The liver is 5 cm BCM with a span of 8 cm.
BP: Right upper arm: 90/50, left leg 60/40
CXR: cardiomegaly with pulmonary venous congestion

ECG: Right ventricle hypertrophy (can be normal for age)

ABG: PH: 7.22, PCO2: 25, PO2: 70, HCO3 10, BE: -15 (metabolic acidosis).
Diagnosis: Severe neonatal coarctation
Definitive diagnosis:
-Echo
-CT scan

Differential diagnoses:

1. Critical aortic stenosis.
2. Hypoplastic left heart syndrome (HLHS).

Noncardiac diagnoses:

- Severe sepsis.
- Metabolic disorder.

Management:

- Supportive: **A**irway, **B**reathing, **C**irculation
- Prostaglandin E1 infusion: as above
- Diuretics: Furosemide
- Dopamine and dobutamine infusions may be needed

Surgical Repair:

- Coarctation repair: needs to be done urgently.
- Critical AS: treatment depends on the size and morphology of the aortic valve, the presence of AR and associated hypoplasia

of the aortic arch and the size of the mitral valve and left ventricle. Balloon valvoplasty or surgical valvotomy can be done with comparable results but there is a high rate of re intervention.

- HLHS needs staged palliation in experienced centres (Norwood procedure). The outcome of this 3-stage procedure is improving attributed to an increased surgical and interventional catheterization experience. When the anatomy is not favourable for palliation, heart transplant would be the only option. This is an extensive procedure which is done in selected centres, however the results have been improving after introduction of new immunosuppressive drugs like mycophenolate mofetil. The early mortality is about 6%, mainly due to graft rejection and the late mortality is around 12%, 5-year survival is 80%.

Scenario B

Collapsed Neonate with severe respiratory distress:

A 12 hour old baby presented with severe respiratory distress.

P/E

HR180, RR 80, Sp02 80 % in room air and 85% on O2. Chest: bilateral crepitations. CVS: No murmur. Liver is not enlarged. Pulses are felt all over.

CXR: Figure 49

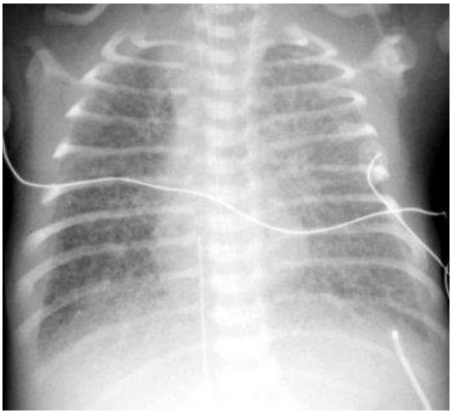


Figure 49
CXR from a patient with severe respiratory distress showing a ground glass appearance and a small heart

Differential diagnoses:

1. Obstructed total anomalous pulmonary venous drainage (TAPVD)
2. Severe pneumonia (congenital)
3. Meconium aspiration
4. Respiratory distress syndrome

Echo revealed that all 4 pulmonary veins drain to the inferior vena cava below the diaphragm with obstruction at the inferior vena cava, gradient of 15 mmhg.

Final diagnosis: Obstructed TAPVD

Management of obstructed TAPVD:

It is a surgical emergency. Supportive treatment and diuretics are needed till surgery is performed. Clinical and CXR appearance can mislead the physician therefore a high index of suspicion should be present. This type of TAPVD is usually infra-cardiac to the inferior vena cava leading to obstruction of the pulmonary veins by the diaphragm. Supracardiac TAPVD to the superior vena cava / innominate vein with a restrictive ASD/PFO can lead to a similar picture in infants.

Scenario C

Neonate with severe cyanosis and heart failure:

A baby was delivered by normal spontaneous vaginal delivery and 6 hours after birth found to have cyanosis and respiratory distress.
P/E
SpO2: 60% in room air and 70% on O2. RR=80/min
No murmurs heard. Liver 3 cm BCM with a span of 6 cm.
Normal pulses

CXR: Egg on side: cardiomegaly, narrow cardiac pedicle, plethoric lungs (see Fig 12)
ECG: Normal neonatal RV dominance.
Echo: TGA with intact interventricular septum.

Differential Diagnosis:
-Severe respiratory disease: normal echo with abnormal CXR.
-Persistent pulmonary hypertension of the newborn: Echo can help to measure the pulmonary pressure from the TR gradient.

Management of TGA:

- Immediate PGE1infusion
- Diuretics
- Urgent referral for balloon atrial septostomy
- Surgical correction.

Discussion:
TGA with intact ventricular septum is an emergency. Early diagnosis and early use of PGE1 are vital. If surgery is

immediately available there is no need to do septostomy. In the first few weeks of life the preferred surgery is the arterial switch operation (resecting and transposing the great arteries with coronary transplantation into the new aorta), which, in experienced hands has a good long term outcome as there is anatomical and physiological correction. The main early complications are related to coronary relocation which can result in mortality related to myocardial infarction. If the patient with TGA presented late the LV would be involuted i.e. is small in volume and thickness because it is connected to the low pressure pulmonary artery. In this case the arterial switch cannot be done unless the LV is initially ‘trained’ by a PA band. The other alternative is to perform atrial switch procedures like Senning and Mustard operations (switching the venous flow at the level of the atria so as to direct the systemic venous flow to the LV then PA and pulmonary venous flow to the RV then aorta). Although this operation results in *physiologic correction* it is associated with the following complications:

1. Systemic/ pulmonary venous obstruction,.
2. Absence of sinus rhythm leading to arrhythmias and occasional sudden death.
3. Late complications are similar to congenitally corrected (L)TGA which are tricuspid valve insufficiency and RV (i.e. systemic ventricle) dysfunction.

Scenario D

The cyanosed Neonate without heart failure: presentation and management similar to scenario 2.

Suggested Readings:

1.de-Wahl Granelli A, Wennergren M, Sandberg K, Mellander M.et al. Impact of pulse oximetry screening on the detection of duct dependent congenital heart disease: a Swedish prospective screening study in 39,821 newborns. BMJ. 2009;338:a3037.

2.Shivananda S, Kirsh J, Whyte HE, Muthalally K.et al. Accuracy of clinical diagnosis and decision to commence intravenous prostaglandin E1 in neonates presenting with hypoxemia in a transport setting. J Crit Care. 2010;25:174.

3.Delmo Walter EM, Hübler M, Alexi-Meskishvili V. et al. Staged surgical palliation in hypoplastic left heart syndrome and its variants. J Card Surg. 2009;24:383-91.

4.Groetzner J, Reichart B, Roemer U, Reichel S.et al. Cardiac transplantation in pediatric patients: fifteen-year experience of a single center. Ann Thorac Surg. 2005 ;79:53-60.

5. A Sound in My Child's Heart (murmurs)

Scenario A

A 6 year old boy was referred from the school health physician because of a heart murmur. He is asymptomatic. Parents were extremely worried.

P/E

Normal looking child, normal vital signs. CVS: Normal S1 and S2, musical blowing ESM 2/6 between the left lower sterna border and apex**.**

Normal CXR, ECG and Echo.

Diagnosis: Innocent “Still’s” murmur

Parents were reassured.

Discussion:

Innocent Murmurs:

Innocent murmurs are a common cause for referral to cardiologist. It is very important to explain to parents that the sound in their child's heart is likely to be innocent unless the suspicion of pathological murmur is strong (site and intensity of the murmur suggestive of pathological murmur or other signs like cyanosis and heart failure).

- Still's murmur: a blowing soft short ESM 1-2/6 in intensity heard between the left sternal border and the apex in children 3-10 years of age.
- Pulmonary flow murmur: soft 1/6 ESM at the pulmonary area in children.
- Physiologic pulmonary artery branch stenosis: soft 1/6 ESM heard at pulmonary area and radiates to the back in neonates up to 6 months.

- Venous hum: heard at the aortic area and the neck, disappears on neck flexion and compression of jugular veins.

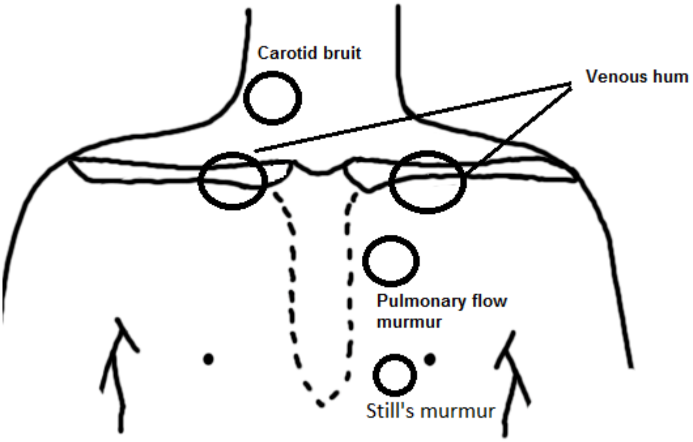


Figure 50
Diagram showing the sites of common innocent murmurs

Scenario B
Murmurs as presenting problems:

An asymptomatic 7 year old child was referred by the family physician because of a murmur heard during routine examination.

P/E: Well looking, normal vital signs, O2 saturation 98%. Normal S1 and S2, a harsh ESM 4/6 (with a thrill) and ejection click are heard at the pulmonary area.

CXR: Normal
ECG: Figure 51

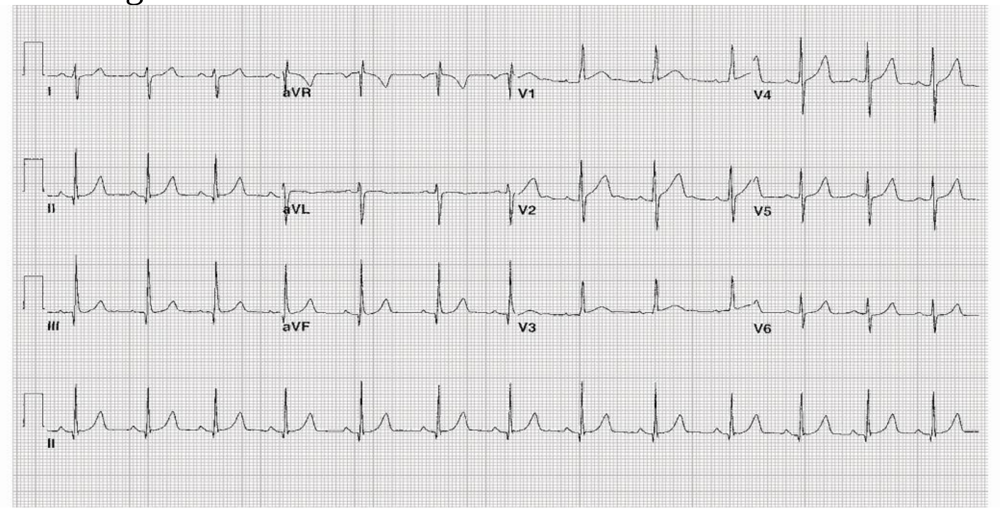


Figure 51
ECG of a 7 year old patient showing right axis deviation and pressure overload RVH (pure R wave, upright T wave in V1)

Echo: Severe pulmonary valve stenosis with a gradient of 90mmHg.
Management:

The patient underwent transcatheter pulmonary valve dilatation. After the procedure Echo revealed a gradient of only 30mmHg across the valve. Regular follow up showed no increase in PV gradient.

Differential diagnosis:

- ESM at the pulmonary area are common. PS murmur depends on the severity of the stenosis. If the stenosis is mild (echo gradient 15-40 mmhg) the murmur is soft and short and can be similar to that of innocent pulmonary flow murmur. With more severe stenosis the murmur is longer and harsher. A click differentiates pulmonary stenosis from other causes (see below).
- Soft ESM at the pulmonary area: Can be due to ASD secundum or primum or *partial* APVD (in TAPVD the patient is symptomatic). Wide fixed splitting is an important sign of ASDs and in APVD. In large ASDs there is an associated tricuspid diastolic flow murmur.
- ESM at 3rd intercostal space: this is the second aortic area, systolic murmurs can be due to subaortic stenosis (e.g subaortic membrane) or HOCM. Aortic (and pulmonary) regurgitation are heard at this area as a “to and fro” murmur.
- Harsh pansystolic murmur at the 4th intercostal space: small-moderate VSDs are the commonest cause of this murmur. Tricuspid regurgitation (primary or secondary to pulmonary hypertension) can cause of a similar murmur.

Other pathological murmurs: Figure 52

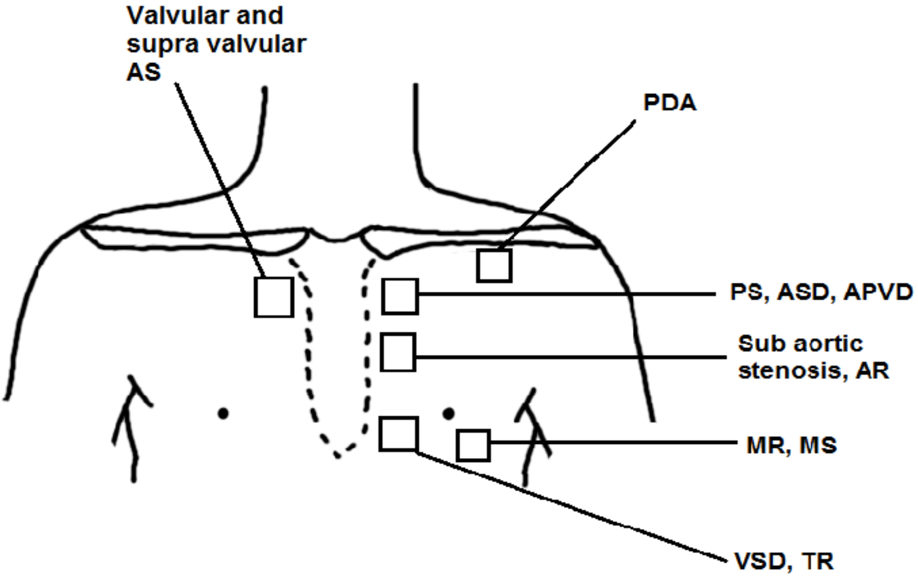


Figure 52
Diagram showing the positions of common pathological murmurs.

6. Eisenmenger's Syndrome

Eisenmenger's syndrome (ES) refers to irreversible pulmonary vascular obstructive disease secondary to untreated large left to right shunts. ES usually occurs in older children/adolescents but the exact age is quite variable. Although clinical and pathological alterations are similar to primary PHT, adults with Eisenmenger's syndrome exhibit increased survival and more favourable hemodynamics than those with idiopathic PHT. In syndromic patients especially Down's syndrome ES can develop as early as few months of age and even at birth.

The sequence of events in patients with untreated large left to right shunts:

1. Age 2 – 24 months: symptoms of CHF, failure to thrive, recurrent infections. Clinical examination: signs of CHF, displaced apex, loud murmurs.CXR: Biventricluar enlargement , atrial enlargement and plethoric lungs. ECG: Biventricular and left atrial enlargement.(As in scenario1).
2. Age 2-7 years (the **'honeymoon period'** symptoms and signs of heart failure disappear, improved weight gain as in scenario 5-A below.
3. After the age of 7 years to adulthood: Typical Eisenmenger's syndrome as in scenario 5- B below.

Differential Cyanosis:

Refers to cyanosis and clubbing in the toes without involvement of the fingers. It occurs in Eismnmenger's syndrome due to a large PDA where the right to left shunt across the PDA causes desaturated blood to supply the lower limbs. (Figure 53). It also occurs in neonates with severe CoA where the PDA shunts from right to left.

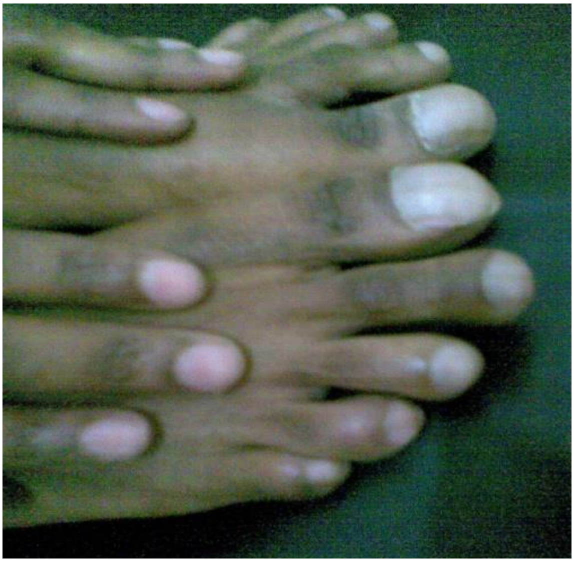


Figure 53
A 20 year old lady with a large PDA. There is clubbing of the toes with normal fingers.O2 sat on the hand is 95% and on the feet 85% (differential cyanosis)

Scenario A

A 3 year old was diagnosed as VSD in infancy when he used to have respiratory distress and failure to thrive. The mother is happy that all these symptoms disappeared in the last 6 month and he is now well. She was advised by her physician to consult the cardiologist to make sure that the VSD has closed as she presumed.

P/E

> Well childe not in failure, no obvious cyanosis or clubbing. O2 saturation 92%. Apex beat at the 5th intercostal space lateral to the mid clavicular line, RV heave is present. A short ESM was heard at LLSB, loud P2.

CXR: The cardio thoracic ratio is 55%, prominent pulmonary artery. Increased pulmonary vascular markings are still present.

ECG: Biventricular enlargement and left atrial enlargement.

Echo: Large perimembranous inlet VSD 10 mm with bidirectional shunt, dilated left side. Mild TR with gradient 70mmhg (PA systolic pressure 80mmHg).

Cardiac Catheterization:

Site	Pressure (mean)	O2 sat % (room air)	Sat on O2
Superior vena cava	-	60	75
PA	75	80	95
Ao	90	92	100
Pulmonary vein	-	95	100

Calculated left to right shunt in room air:

QP/QS = AoSat- SVC sat/ Pul vein- PA sat
QP/QS in room air = 92-60/95-80
= 2.1:1
QP/QS on 100% O2 = 3.6:1

Calculated pulmonary vascular resistance in room air =10 Woods units and on O2 dropped to 8 Woods units.

Diagnosis: Large VSD with reversible (but significantly high) pulmonary HTN

Management:

The patient was discussed with the cardiac surgeon. A high risk VSD closure was done. The post operative course was complicated with pulmonary hypertensive crisis which was managed by nitic oxide then Sildenafil. The patient recovered and was discharged.

Scenario B

A 15 year old boy presented with shortness of breathing on minimal exertion. He had history of CHD and was very symptomatic as a baby then symptoms disappeared for many years and the family was reassured by the physician. Recently he developed easy fatigability.

P/E

Stunted growth. O2 sat 85%. There is cyanosis and clubbing. A short ESM heard at LLSB. Loud P2, No signs of heart failure.

CXR

Small heart with right ventricle configuration. Prominent main pulmonary artery and central branches with sudden tapering and oligemia peripherally (pruning, see figure 8)

ECG: Right axis deviation, RVH. No LVH

Differential diagnoses:

1. Eisenmenger's syndrome
2. Pulmonary hypertension: Primary or secondary to noncardiac causes.

Echo:

A large perimembraous inlet VSD 12 mm with bidirectional shunt. The right sided is dilated. Normal left ventricle and left atrial dimensions.

Cardiac cath:

Cardiac Catheterization:

Site	Pressure (mean)	O2 sat %(room air)	sat on O2
Superior vena cava	-	60	70
PA	90	70	80
Ao	90	85	90
Pulmonary vein	-	95	100

Calculated left to right shunt in room air:

QP/QS = AoSat- SVC sat/ Pul vein- PA sat

QP/QS in room air = 1:1

QP/QS on 100% O2 = 1.4:1

Calculated pulmonary vascular resistance in room air =26 Woods units and on O2 23Woods units.

Diagnosis: Large VSD with *irreversible* pulmonary vascular obstructive disease (Eisenmenger's syndrome)

Management:

The patient is inoperable. He was started on sildenafil with some symptomatic improvement.

Discussion:

Management of Eisenmengers syndrome:

1. Surgical closure of the shunt is contraindicated in well-established cases.
2. Treatment of right sided heart failure with diuretics, modest doses should be used.
3. Regular check for haemoglobin and hematocrit levels, to keep Hb >12 gm/dl with normal MCV and MCH.

4. Indications for partial exchange transfusion : In asymptomatic patients a hematocrit > 68% and in symptomatic patients (headache, fatigue, blurring of vision) exchange transfusion is indicated for hematocrit >60%. Always give normal saline when vensection is done to avoid increasing the blood viscosity.
5. Pulmonary vasodilators: different drugs had been tried like prostacyclins and bosentan (endothelin receptor antagonist). Bosentan proved to be effective in improving the functional class in patients with primary PHTN and Eisenmenger’s syndrome; however both prostacyclins and bosentan are quite expensive. Sildenafil (phosphodiesterase 5 antagonist) is a relatively cheap and effective pulmonary vasodilator that had been used widely to treat patients with primary pulmonary hypertension and ES with good results. There have been some case reports about the surgical closure of ASD in patients with Eisenmenger’s syndrome after prolonged treatment with Sildenafil.
6. Heart- lung (or lung only) transplant is the only definitive treatment for ES.
7. Early detection and referral of patients with CHD can prevent this devastating complication.

Suggested Readings

1.Tay EL, Papaphylactou M, Diller GP, Alonso-Gonzalez R, Inuzuka R, Giannakoulas G, Harries C, Wort SJ, Swan L, Dimopoulos K, Gatzoulis MA. Quality of life and functional capacity can be improved in patients with Eisenmenger syndrome with oral sildenafil therapy. Int J Cardiol. 2010 Mar 19. [Epub ahead of print]

2.Waddell TK, Bennett L, Kennedy R, Todd TR, Keshavjee SH.Heart-lung or lung transplantation for Eisenmenger syndrome. J Heart Lung Transplant. 2002 Jul;21(7):731-7.

3.Diller GP, Dimopoulos K, Kaya MG, Harries C, Uebing A, Li W, Koltsida E, Gibbs JS, Gatzoulis MA.Long-term safety, tolerability and efficacy of bosentan in adults with pulmonary arterial hypertension associated with congenital heart disease. Heart. 2007 Aug;93(8):974-6.

4.Kim YH, Yu JJ, Yun TJ, Lee Y, Kim YB, Choi HS, Jhang WK, Shin HJ, Park JJ, Seo DM, Ko JK, Park ISRepair of atrial septal defect with Eisenmenger syndrome after long-term sildenafil therapy. Ann Thorac Surg. 2010 May; 89(5):1629-30.

5.Sulafa KM Ali Experience with Sildenafil in treatment of pulmonary hypertension in Sudan. Sudanese Journal of Paediatrics.2007;8:91-98

7. Acute Rheumatic Fever and Rheumatic Heart Disease

Scenario 1

New episode of ARF with arthritis and carditis

A 7 year old boy presented with migratory arthritis of the knee and ankle joints and shortness of breath for 7 days. The joint pain and swelling continued for 3 days then subsided to affect another joint.

P/E

HR 150 /min, RR 50/min, temp 38 degrees,

CVS: The apex beat is in the 6th intercostals space lateral to the mid clavicular line, heaving in nature. There was a palpable thrill and positive right parasternal heave. A pansystolic apical murmur with mid diastolic murmur were heard. A tender liver 4 cm BCM with a span of 9 cm was palpable.

CXR:LV and LA enlargement

ECG: Figure 54

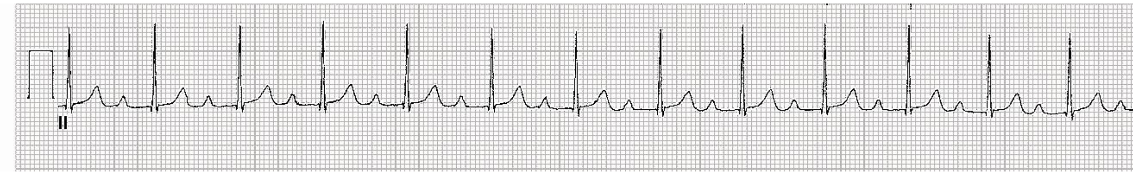


Figure 54:

ECG of a patient with ARF showing first degree AV block (PR interval=0.32 seconds)

Other investigations:

Antistreptolysin O titre > 200, ESR is 100mm/hour.

Echo: severe MR, mild AR, small pericardial effusion.

Diagnosis:

Acute rheumatic fever (ARF). The patient is fulfilling the modified Jone's Criteria: the 2 major criteria are migratory arthritis and carditis and the 3 minor criteria are fever, high ESR and long PR interval plus evidence of streptococcal infection (positive ASO titre).

Differential Diagnosis:

Connective tissue disease (SLE or juvenile idiopathic arthritis) should be considered if the pattern of arthritis is non migratory, involving large and small joints or more than 2 weeks in duration.

Management:

1. Bed rest.
2. Penicillin : one dose of benzathine penicillin: 600000 IU for patients <30 kg and 1.2 million IU for those >30 kg.
3. Anti inflammatory drugs: aspirin 75 mg/kg/day for 2 weeks then taper as ESR comes down.
4. 3 weekly benzathine penicillin prophylaxes, same dose as above should be continued for life if there is residual valve lesion.
5. Anti heart failure treatment: Furosemide and captopril.

Scenario 2

Recurrence of ARF with carditis

The same patient in scenario 1 went home then lost to follow up for 18 months, he used penicillin only for the first 3 month, this time he came complaining of shortness of breathing and arthralgia of the knee and hip joints

P/E

Distressed, HR 160/min, large volume pulse, RR 5O/min, temp 38. CVS: apex at 7th intercostals space outside the midclavicular

line, heaving in character. Pansystolic apical murmur, mid diastolic apical murmur. A to-and –fro (systolic-early diastolic) murmur is heard at the second aortic area. Loud P2
Liver 4 cm BCM (span 9 cm), tender.

Differential diagnoses:

1. Recurrence of acute RF with carditis, MRand AR, no vegetations.
2. Rheumatic heart disease (RHD): MR and AR with heart failure and fever for investigations (endocarditis or other causes of fever).

Investigations:
ESR: 120mm/hour, ASO titre>200
Echo: Severe rheumatic MR and AR
CBC: normal, blood culture: negative
Diagnosis:
Recurrence of ARF.
This diagnosis is based on 3 minor criteria (arthralgia, fever and high ESR) with evidence of streptococcal infection. Recurrence is commonly associated with carditis so minor criteria and evidence of streptococcal infection should be carefully looked for in such patients.
Treatment:
The same as a new episode of ARF. In severe AR it is better to avoid angiotensin converting enzyme inhibitors as they may lead to exaggeration of hypotension.

Scenario 3
Established rheumatic heart disease (RHD)

A 14 year old boy presented with shortness of breathing and palpitations on mild exertion for 6 months, no history of joint pain or fever.
P/E

Ill, a febrile. RR 40 breaths/min, HR 120 beats/min, irregular. Apex beat at the 5th intercostals space within the mid clavicular line, tapping in character. Right ventricle heave and palpable P2 were found. Apical mid diastolic rumbling murmur. Loud P2.

Chest: fine basal crepitations.

Abdomen: The liver is 4 cm below the costal margin with a span of 9 cm, tender.

ASO: <200, Normal CBC and ESR

CXR: Left atrial enlargement, pulmonary edema.

ECG: Figure 55

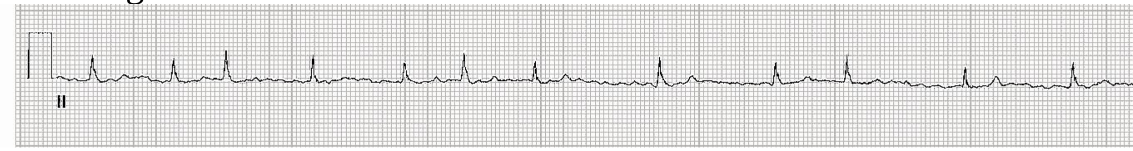


Figure 55

ECG of a patient with mitral stenosis showing atrial fibrillation: the P wave is not seen, fibrillation waves with irregular QRS.

Echo:

Severe mitral stenosis, no MR. Moderate tricuspid regurgitation with high pulmonary artery pressure.(Figure 56)

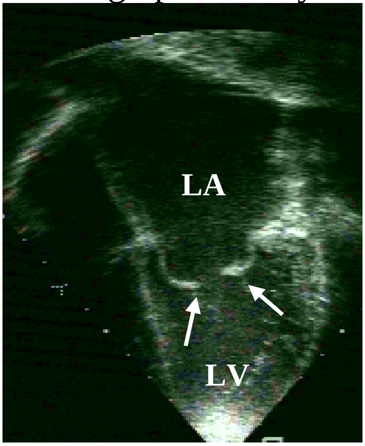


Figure 56 (above): Echocardiographic 4 chamber view showing thickened retracted mitral valve leaflets (arrows) with restricted opening. Dilated left atrium (LA) and normal size LV in a patient with rheumatic MS.

Diagnosis:

Established rheumatic MS with atrial fibrillation and pulmonary hypertension.

Management:

i. Diuretics
ii. Aspirin: antiplatelet dose
iii. Digoxin
iv. Trans-catheter balloon mitral commissurotomy.

Discussion:

- ARF and its devastating sequel RHD are a major public health problem in the Sudan. It is due to infection by group A beta hemolytic streptococci (GABS) with consequent immunological host interactions leading to joint, heart, skin and nervous system affection. All the manifestations disappear except the valvular affection which leads to permanent damage.
- The diagnosis of ARF is based on the Jone's Criteria: the major criteria are: arthritis, carditis, chorea, erythema marginatum and skin nodule. The minor criteria are fever, arthralgia, increased acute phase reactants and prolonged PR interval on the ECG. The diagnosis of a new case is based on 2 major or 1 major and two minor criteria plus evidence of acute streptococcal infection (positive antistreptolysin O or positive throat culture).
- The Jones Criteria were proposed in 1944 and remained the gold standard for diagnosis. The current criteria were

revised in 1992 by the American Heart Association and in 2003 by the World Health Organization. Table 12 shows the current WHO criteria.

- The most important modification in the Jones Criteria is the special consideration to patients with recurrence of ARF. There should be a low threshold to diagnose recurrence as it carries a high risk of cardiac involvement. *Two minor* criteria plus evidence of streptococcal infection is considered diagnostic of recurrence.
- Diagnosis of carditis is clinical (CHF, murmurs suggestive of RHD). The apical mid diastolic murmur in ARF is called the Carry Coombs murmur, it is related to valvulitis, however severe MR whether acute or chronic is also associated with a functional mid diastolic apical murmur.

The Role of Echo In Assessment of RHD:

- Echo is cruicial in evaluation of cardiac involvement in ARF and RHD,it should be performed using stgandard methodology and reference values.
- Objective echo assessment of MR is measurement of the jet characteristics(width of MR jet, jet: LA area ratio, flow reversal in pulmonary veins, the size of LA and LV, measurement of pulmona artery pressure and the ejection fraction,
- AR is assessed by measurement of the jet characteristics (jet width and the jet: LV outflow ratio), AR pressure half time and flow reversal in the descending aorta. LV size and function as for MR.
- Leaflet thickening is commonly encountered in RHD however there no published standard reference values for normal and abnormal leaflets thickness. We recently found that the normal (mean) anterior leaflet thickness is 3 mm/m2 compared with 6.3 mm/m2 in patients with severe MR while the thickness of posterior mitral leaflet in

normal children was found to be 2.5 mm/m2 compared with 5.7 mm/m2 in patients with severe MR.

- So far echo is not included as a diagnostic criterion for ARF but patients who show echocardiographic valve regurgitation without clinical signs of carditis should be carefully followed up to determine the outcome of this so called ‘subclinical carditis’.
- Over interpretation of physiological or trivial valvular regurgitation may result in over diagnosis of RHD.

The role of steroids:

There is currently no evidence that steroids should be used routinely for rheumatic carditis. Therefore it should be kept for patients who could not tolerate or did not respond to aspirin and those who are severely ill.

Prevention of ARF:

Primary prevention:

-Rrefers to treatment of GABS pharyngitis with antibiotics to prevent subsequent ARF.

-Primary prevention was proved by many studies to be cost effective in controlling ARF and RHD in many countries where the disease is prevalent. The clinical diagnosis of streptococcal pharyngitis is based on the findings of sore throat, pharyngeal exudates with or without cervical lymphadenopathy and fever in the absence of symptoms of runny nose. As bacteriological documentation is not practical in developing countries, these constellations of symptoms and signs are taken as evidence of bacterial pharyngitis and antibiotics are given. Oral penicillinfor 10 days can be used however one dose of intramuscular benzathine penicillin is more effective. Having a low threshold for diagnosing and treating bacterial pharyngitis is the cornerstone of primary prevention which, with consolidation of public awareness and improvement of socio-economic environment, was shown to decrease the prevalence of this disease.

Secondary prevention:

Referes to the regular use of antibiotics in the form of 3 weekly benzathine penicillin in patients who already developed an episode of ARF. It should be continued for life in patients with RHD. In patients without carditis it is recommended to continue prophylaxis for 5 years after the last attack, or until 18 years of age (whichever is longer).

- There is continuous laboratory research to develop a vaccine against GABS, however the results are not yet ready to be tested in large scale clinical studies.

Differntial Diagnosis of Valvular Heart Disease:

Mitral Valve Disease:

- Differneial diagnosis of MR includes:
    - Congenital MR: Due to dysplastic mitral vavle leaflets. Usually presents with severe CHF in infants and toddlers with a progressive course.
    - Mitral valve prolapsed disease: usually presents in adolescents and young adults with slow progression in childhood. It can be associated with ASD and accessory patheway re entry SVT.
    - Other congenital MV defects include mitral valve cleft (which can be isolated or associated with other CHD) and double orifice MV.
- Mitral stenosis: congenital MS can result from anomalies involving the valve leaflets and papillary muscles (e.g.single papillary muscle leading to parachute MV).

Supra MV rings can lead to MS usually in toddlers and older children.

Aortic valve disease:

- Bicuspid AV: A common cause of aortic stenosis and /or regurgitation. The valve dysfunction usually presents in older children. Severe forms can present in the neonte (critical AS)
- AR can result from prolapse of the AV cusp into small VSD leading to progressive regurgitation.

Pulmonary and tricuspid valve disease:

- Primary diseases of pulmonary and tricuspid valves are uncommon. Ebstein anomaly was discussed in scenario 3.
- Primary pulmonary regurgitation rarely occurs due to ‘absent pulmonary valve syndrome’

Connective Tissue diseases can lead to MR/MS, AR/AS.

Surgical Management of RHD:

Patients with severe valve lesions in functional class III-VI or showing deterioration of EF and LV size on echo should be refereed early for surgery. Valve repair is feasible in experienced centres and is the treatment of choice; however it is often technically challenging especially in patients with combined MR/MS and those with AR. Currently monst patients with RHD are treated with valve replacement which is considered an additional burden on these children and their families.

Problems associated with Valve Replacement:

1. Life-long anticoagulation: patients need to be on regular warfarine for life aiming at an INR of 2.5-3.5. Complicaions related to bleeding can be fatal.On the other hand a sub-optimal INR can lead to valve thrombosis which can lead to serious complications.

2. Patients are at a high risk for endocarditis therefore good dental health and strict endocarditis prophylaxis need to be emphasized.

3. Life- long follow up is needed for theses children whose families are coming from low socioeconomic background where compliance with treatment and follow up is not optimal.

Table 12

2002–2003 WHO criteria for the diagnosis of rheumatic fever and rheumatic heart disease

Diagnostic categories	**Criteria**
Primary episode of RF	Two major or one major and two minor manifestations **plus** evidence of a preceding group A streptococc infection
Recurrent attack of RF in a patient **without** established rheumatic heart disease.	Two major or one major and two minor manifestations **plus** evidence of a preceding group A streptococcal infection.
Recurrent attack of RF in a patient **with** established rheumatic heart disease	Two minor manifestations **plus** evidence of a preceding group A streptococcal infection.
Rheumatic chorea	Other major manifestations or evidence of group A streptococcal infection not required.
Chronic valve lesions of RHD presenting for the first time with pure mitral stenosis or mixed mitral valve disease and/or aortic valve disease.	Patients Do not require any other criteria to be diagnosed as having rheumatic heart disease.

Suggested Readings

1.Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. JAMA. 1992;268:2069-73.

2.Rheumatic Fever and Rheumatic Heart Disease, Report of a WHO Expert Consultation, Geneva, World Health Organization (Technical Report Series, No.923) 2001.

3.Ozkutlu S, Ayabakan C, Saraçlar M.Can subclinical valvitis detected by echocardiography be accepted as evidence of carditis in the diagnosis of acute rheumatic fever? Cardiol Young. 2001 May;11(3):255-60.

4.Cilliers AM, Manyemba J, Saloojee H. Anti-inflammatory treatment for carditis in acute rheumatic fever. Cochrane Database Syst Rev. 2003;(2):CD003176.

5.Soudarssanane MB, Karthigeyan M, Mahalakshmy T, et.al. Rheumatic fever and rheumatic heart disease: primary prevention is the cost effective option. Indian J Pediatr.,74:567-70,2007.

6. Colquhoun SM, Carapetis JR, Kado JH et.al. Rheumatic heart disease and its control in the Pacific. Expert Rev Cardiovasc Ther.,7:1517-24,2009.

7. Robertson KA, Volmink JA, Mayosi BM. Antibiotics for the primary prevention of acute rheumatic fever: a meta-analysis. BMC Cardiovasc Disord 2005; 5:11.

8.Guilherme L, Postol E, Freschi de Barros S, Higa F, Alencar R, Lastre M, Zayas C, Puschel CR, Silva WR, Sa-Rocha LC, Sa-Rocha VM, Pérez O, Kalil J.A vaccine against S. pyogenes: Design and experimental immune response. Methods. 2009;49:316-21.

9.Ganesan Karthikeyan, Bongani M. Mayosi, Is Primary Prevention of Rheumatic Fever the Missing Link in the Control of Rheumatic Heart Disease in Africa? *Circulation*. 2009;120:709-713.)

8.Myocarditis and Cardiomyopathy

Scenario 1
Acute Heart Failure

A 6 months old girl presented with fever and shortness of breathing for 5 days.
P/E Pale, ill, drowsy. HR 170 beats/min, small volume pulse, RR 60 breaths/min, temp 38.5 degrees, weight 8 kg. O2 sat 90% in room air and 98% on O2/ . BP 40/20
CVS: Apex displaced laterally, gallop rhythm, soft SEM at apex
Liver: 5 cm below the costal margin with a span of 9 cm, tender

CXR: Cardiomegaly, LV and left atrium with pulmonary oedema
ECG: low voltage QRS

Differential Diagnosis:

1. Acute myocarditis (viral)
2. Anemic heart failure can be considered from the clinical picture

NB: Pallor is a sign of heart failure due to peripheral vasoconstriction (sympathetic stimulation) with a normal hemoglobin level.

Echo: dilated left ventricle, poor LV function: ejection fraction 20%. normal anatomy. Mild MR.(Figure56)

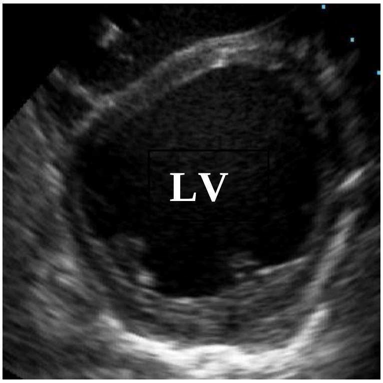


Figure57: Echocardiographic short axis view showing dilated LV with diastolic dimension of 4 cm in a 6 month old infant with acute myocarditis.

Diagnosis: acute myocarditis

Management: the patient should be admitted to ICU with close monitoring.
Supportive treatment:
ABC: Airway, Breathing, Circulation
Investigations:

- Complet blood counts
- Urea, creatinine and electrolytes
- Serum Calcium.
- Viral antibody titres (acute and convalescent) for common viruses like Coxaki B virus.

Specific Treatment:

1. To increase the BP use Dopamine 8-12 mics/kg/min **combined** with Dobutamine 5-10mics/kg/min. Dopamine increases the BP and contractility but in high doses (> 6microgram/kg/min) causes peripheral vasoconstriction which has a deleterious effect on the myocardial function as well as other organs' perfusion therefore. Dopamine in

lower doses will not increase BP but has a mild vasodilatation effect. Dobutamine acts as an ino-dilator so helps the myocardial contractility as well as afterload reduction; overcoming the vasoconstriction caused by high dose dopamine**.**

2. These medications should be titrated according to the response of the patient that is monitored in the intensive care unit by invasive and non invasive BP and central venous pressure measurements. When the cardiac output improves, i.e. urine output, level of consciousness, skin temperature and lastly BP, withdrawal of inotropic drugs should be done gradually.
3. Diuretics: furosemide 2-3 mg/kg/d
4. Consider using intravenous immunoglobulin infusion: its use has not been proven to improve the outcome, however it can be tried in severely ill patients with acute onset of myocardits as in this scenario.
5. On withdrawal of inotropes, angiotensin converting enzyme inhibitors and digoxin can be started.
6. Follow up by clinical and echo examination.

Scenario 2
Chronic heart failure without murmurs

A 2 year old boy came to the emergency room with shortness of breathing, this had been recurrent over the last 6 month and labeled as 'chest infections'

P/E

Ill , wasted, HR 150 beats/min, RR 50 breaths/min, BP 75/50. Weight 7 kg. Chest deformity (Harrison's sulci). The apex is displaced laterally.

Muffled heart sounds, soft systolic murmur at the apex.

CXR: Cardiomegaly, LV and LA enlargement, pulmonary oedema
ECG: Left ventricle hypertrophy, T/ST changes

Echo: same as scenario 1

Diagnosis: Dilated CMP
Treatment

1. Diuretics
2. ACE inhibitors: use small doses with BP monitoring
3. Digoxin
4. Betablockers: carvedolol was proved by large scale studies to be effective in adults with CHF. In the paediatric age many studies had shown clinical and echocardiographic improvement with the use of carvedolol, however the results cannot be compared with adults because of the limited number of patients. It is generally used as a 4th line drug when patients are not responding to conventional treatment.
5. Aspirin as prophylaxis against ventricular thrombosis.
6. Nutritional support

Further options: DCM is a disease with poor prognosis; many new therapeutic options are available in highly specialized centres.

- Cardiac resynchronization therapy: in patients with severe LV dysfunction and prolonged QRS duration (QRS duration >120 msec) biventricular pacing can improve the myocardial function. This treatment had been tried in children in small trials which had shown encouraging results.
- Left ventricle assist devices: internal or external pumps connected with the left ventricle. They are used as a

bridge to heart transplant, however some patients with acute myocarditis can improve on these devices.

- Extracorporeal Membrane Oxygenator (ECMO): using large cannulae, blood is diverted from the caval veins into an external oxygenator then pumped back to the ascending aorta. It is used as a bridge to heart transplant, rarely myocardial recovery can occur.
- Heart transplant is the only curative treatment for end stage cardiomyopathy.
- Recently stem cell transplantation via the coronary arteries has been described and preliminary results are encouraging.

Discussion:

- Paediatric CMP has a wide range of aetiological factors including genetic, infectious and metabolic causes.
- Many inborn errors of metabolism involving cardiomyopathy exist, including fatty acid oxidation defects, organic acidemias, amino acidopathies, glycogen storage diseases and congenital disorders of glycosylation as well as peroxisomal, mitochondrial, and lysosomal storage disorders. Many of these disorders affect both skeletal as well as cardiac muscles.
- Several lysosomal storage disorders are now treatable by enzyme replacement therapy and/or bone marrow transplantation e.g. the treatment of Pompe’s disease (glycogen storage disease type II) with alpha glucosidase enzyme replacement which has changed the outlook of these patients.

Types of CMP:

Cardiomyopathies are generally classified as dilated (DCM), hypertrophic (HCM), restrictive, primary genetic and ‘unclassified’ types. Echo plays a central role in diagnosis and follow up.

Dilated CMP:

- DCM is the most common type of CMP and constitutes 70% of cases. The aetiology is variable, the most common cause is post-viral myocarditis which develops in about 30% of patients who have acute viral myocarditis.
- Human immunodeficiency virus (HIV) is an important cause of DCM in endemic areas. Decreased myocardial function was found in 35% of patients with HIV. Other echocardiographic findings in HIV include diastolic dysfunction (19%) and pericardial effusion (11%). CD4 cell count <100 cells/mm3 was associated with high prevelance of cardiac abnormalities. With the introduction of antiretroviral treatment many children with HIV lived into adulthood where studies had shown an increased prevelance of cardiovascular risk factors and premature atheroscelrosis.
- Echo cannot differentiate acute myocarditis from chronic cardiomyopathy, in both conditions it will show a dilated left ventricle with poor ejection fraction. The clinical course in acute myocarditis is short with a history of a preceding febrile illness. The patient is well grown with no features of chronic heart failure.
- ECG is helpful; it can show decreased voltage in acute myocarditis in contrast to increased left ventricle forces in cardiomyopathy.
- Viral serology may help in some patients.
- Recently, cardiac magnetic resonance imaging (MRI) proved to have an important role in the diagnosis of acute myocarditis by detection of myocardial edema. Endomyocardial biopsy in specialized centers may differentiate these entities but it also has many limitations.

Important Causes of DCM that needs to be ruled out:

1. **A**nomalous **L**eft **C**oronary from the **P**ulmonary Artery (ALCAPA) Syndrome (also known as Bland-White-Gerland syndrome): is a rare and potentially curable cause of DCM. It usually presents with CHF in infants and echo can be similar to DCM. The ECG can be diagnostic showing deep and wide Q wave seen in I, aVL, and V6 characteristic of anterolateral myocardial infarction.(Figure 58). Careful Echo imaging can show the abnormal left coronary artery which can be confirmed by angiography or MRI. Surgical coronary transfer can be successful when patients are picked up early.

2.Untreated severe CoAo can lead to DCM. Echo should rule out these defects before labelling the patient as DCM.

3.Valvular lesions: severe MR, AR and AS are relatively common causes of secondary DCM. MR should be diagnosed and managed early before myocardial function deteriorates as the myocardial function rarely recovers. On the other hand, surgical correction of AR/AS associated with poor ejection fraction can lead to recovery of myocardial function.

4. Hypocalcemia is an important reversible cause of DCM in infants, serum calcium level should be done in such paients especially those manifesting sings of rickets. DCM was reported as the first sign of rickets in infants.

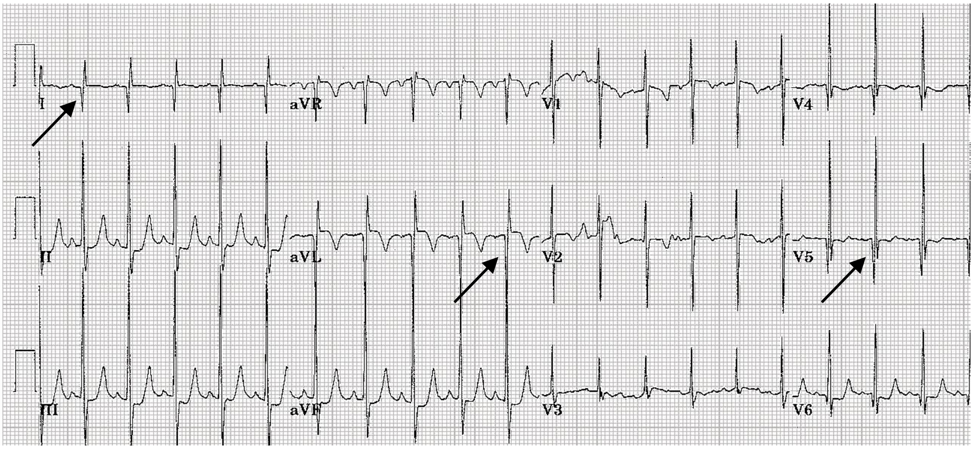


Figure:58
ECG of a 4 months old infant with ALCAPA syndrome showing deep q waves in leads I, aVL,V4-5.(arrows).There is LVH.

Hypertrophic CMP:

Occurs in 10% of patients. The infantile type can be associated with genetic or metabolic disease (e.g. type II glycogen storage disease:Pompe's disease) and Noonan's syndrome. Family history is often positive. The course is progressive with heart failure and failure to thrive causing death in infancy. The adult type (autosomal dominant) occurs in adolescents.

Distinctive Features

Left sided heart failure with pulmonary oedema (wheezing)
Positive family history.

Diagnosis:

Echo finding of hypertrophied left ventricle with or without obstruction to the left ventricle outflow tract (Figure 59). Mitral regurgitation can be present. Systolic function is preserved and usually hyperdynamic while the diastolic function (ventricular relaxation) is impaired.

Management:

1. Diuretics are used in small doses to improve the symptoms of pulmonary edema.

2. Betablockers: Propranolol.
3. Surgery: septal myomectomy is occasionally done for patients with severe left ventricle outflow tract obstruction, it has shown good results in experienced centres with 98% 10- year survival rate.
4.Transcathere septal reduction by alcohol ablation is a less invasive technique with results that are comparable to surgery.

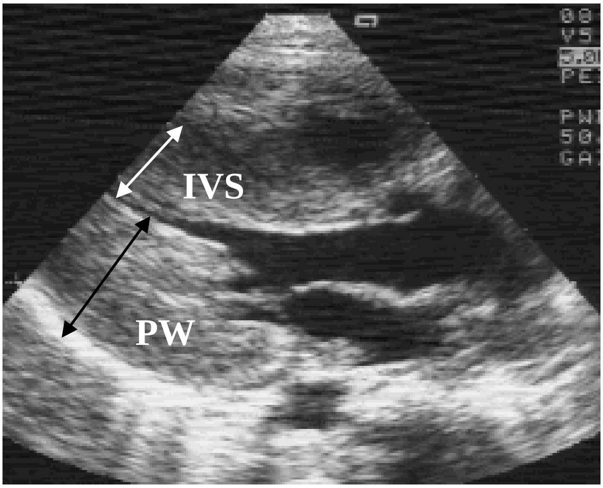


Figure 59:
Echocardiographic parasternal long axis view of a 6 months old showing severe LVH: the interventricular septum (IVS) is 1.5 and LV posterior wall (PW) is 2.5 cm with mid-cavity obstruction.

High risk patients:
Familial history of sudden death at a young age, LVH >3 cm, unexplained syncope, nonsustained ventricular tachycardia on Holter monitoring, and abnormal blood pressure response during exercise are considered risk factors for sudden death. Primary prevention of sudden death in patients considered to be at high risk should aim at the management of obvious arrhythmogenic mechanisms, and the prevention and/or management of

ventricular tachyarrhythmias with amiodarone and/or implantable cardioverter defibrillator (ICD), respectively. The choice of treatment in children is influenced by technical aspects, such as adverse effects of amiodarone, and ICD implantation difficulties or complications. Amiodarone could also be used as a bridge in children at high risk, until they reach adulthood, possibly achieving a lower risk status, or until their physical growth permits ICD implantation as long-term therapy.

Other types of CMP:

Noncompaction CMP:

A primary genetic CMP with variable presentations and constitutes about 9% of all CMP.

Distinctive Features:

- Most patients present with heart failure due to myocardial dysfunction.
- Can be present in asymptomatic individuals.
- Relapses and remissions of myocardial dysfunction.
- Female preponderance in Arab and African patients while it is more common in males in the Western countries.
- Familial tendency
- Association with simple and complex CHD.
- Thromboembolism which is reported in 9% of patients probably related to stagnation of blood in the intertrabecular recesses.
- Association with mitral regurgitation: deformity of mitral leaflets with severe mitral regurgitation can be present *without* left ventricle dysfunction.

Diagnosis:

Echo is diagnostic showing prominent myocardial trabeculations and deep intertrabecular recesses. There is a 2- layer appearance (one compacted and the other is noncompacted) with noncompacted to compacted layer thickness ratio of 2:1or more. (Figure 60)

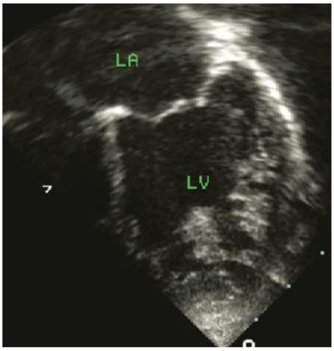


Figure 60:
Echocardiographic modified 4-chamber view showing multiple trabeculations in the LV and abnormal mitral valve leaflets in a patient with noncompaction CMP.

Management:
Diuretics, ACE inhibitors as in other patients with CHF. Aspirin to prevent thrombosis. Patients with MR or CHD may need surgical intervention. The risks of myocardial dysfunction and arrhythmia should be carefully discussed with the family as well as with the surgeon.

Restrictive CMP

This type is rare in children (less than 1%), however the most common type of restrictive CMP world wide is called tropical restrictive CMP or Endomyocardial fibrosis (EMF) which is frequently encountered in Sudan and nearby countries. The disease is related to parasitic infestations particularly with bilharziasis and filariasis.

Clinical Features of EMF

The disease is more common in males 12-15 years of age from poor families. Insidious onset of abdominal distension that can be massive without lower limb edema (Fig 61). Shortness of breathing occurs if there is left side affection (uncommon)

On examination: There is growth delay, prominent ascities, raised jugular venous pressure, hepatosplenomegaly (signs of restrictive CMP and constrictive pericardites). Murmurs are uncommon.

Investigations: Eosinophilia can be found in peripheral blood. Tropical infestations especially bilharziasis should be excluded.

CXR: Cardiomegaly with right atrial enlargement

ECG : is characteristic showing atrial enlargement that can mimic Ebsteins' anomaly with Himalayan P wave (Figure62). Atrial fibrillation and AV block can be present

Echo: Diagnostic features include huge right atrium with spontaneous echo contrast and obliteration of the right ventricle apex by fibrous or calcified tissue (Fig 63). Deformity of the tricuspid valve can cause confusion with Ebstein's disease. Pericardial effusion can be present.

Differential Diagnosis:

Clinical features can mimic constrictive pericarditis (in tropical areas can be related to tuberculosis).

Management:

Diuretics

Digoxin or betablockers for atrial fibrillation

Aspirin

Surgical treatment is not satisfactory. In selected cases cavopulmonary anastomosis can be done to unload the right atrium and ventricle.

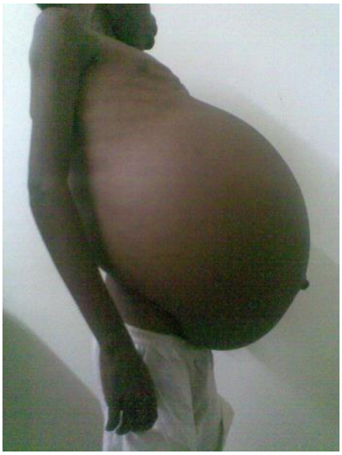


Figure 61:
A patient with endomyocardial fibrosis with massive ascites, note the body wasting

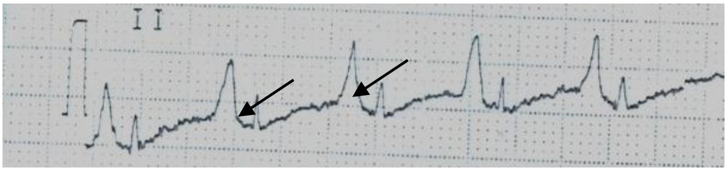


Figure 62:
ECG (lead II) of the patient in Figure 61showing Himalayan P wave that is double the QRS voltage (arrows)

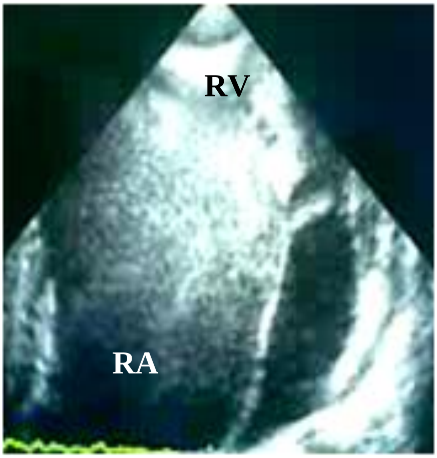


Figure 63
Echo 4 chamber view of a the patient in figure 61 showing massive RA with spontaneous echo contrast and calcification of RV apex.

Suggested Readings

1.Klugman D, Berger JT, Sable CA, He J, Khandelwal SG, Slonim ADPediatric patients hospitalized with myocarditis: a multi-institutional analysis. Pediatr Cardiol. 2010 ;31:222-8.

2.-Myra Cocker & Matthias G. Friedrich. Cardiovascular Magnetic Resonance of MyocarditisCurr Cardiol Rep 2010;12:82–89

3.Bajcetic M, Kokic Nikolic A, Djukic M, Kosutic J et al.Effects of carvedilol on left ventricular function and oxidative stress in infants and children with idiopathic dilated cardiomyopathy: a 12-month, two-center, open-label study. Clin Th 2008 ;30:702-14.

4 Aggarwal P, Sharma A, Bhardwaj R, Raina R. Myocardial dysfunction in human immunodeficiency virus infection: an echocardiographic study.J Assoc Physicians India. 2009 Nov;57:745-6.

5. Sims A, Hadigan C Cardiovascular Complications in Children with HIV Infection.Curr HIV/AIDS Rep. 2011 Jun 4. [Epub ahead of print]

6.Abdullah M, Bigras JL, McCrindle BW. Dilated cardiomyopathy as a first sign of nutritional vitamin D deficiency rickets in infancy.Can J Cardiol. 1999 Jun;15(6):699-701.

7.Läer S, Mir TS, Behn F, Eiselt M et al. Carvedilol therapy in pediatric patients with congestive heart failure: a study investigating clinical and pharmacokinetic parameters. Am Heart J. 2002 ;143:916-22.

8.van der Ploeg AT, Reuser AJ.Pompe's disease. Lancet. 2008;372:1342-53.

9. Bokeriia LA, Borisov KV, Sinev AF.Long-term effects of surgical septal myectomy on survival in patients with obstructive hypertrophic cardiomyopathy. J Am Coll Cardiol. 2005;46:470-6.

10.Alam M, Dokainish H, Lakkis N. Alcohol septal ablation for hypertrophic obstructive cardiomyopathy: a systematic review of published studies. J Interv Cardiol. 2006;19:319-27.

11.Sulafa KM Ali.Unique features of noncompaction of the ventricular myocardium in Arab and African patients. Cardiovas J South Afr 2008;19:241-45.

12.Sulafa KM Ali. Endomyocardial fibrosis: an under diagnosed cause of restrictive cardiomyopathy in Sudanese children. J Trop Paed 2009;10:147-51

13.Anjan S Batra, Seshadri Balaji.Cardiac Resynchronization Therapy in Children. Curr Cardiol Rev. 2009 ; 5: 40–44.

14.Zimmerman H, Covington D, Smith R, Inaht C et al.Recovery of Dilated Cardiomyopathies in Infants and Children Using Left Ventricular Assist Devices. ASAIO J. 2010.

15.Olguntürk R, Kula S, Sucak GT, Ozdoğan ME et al. Peripheric stem cell transplantation in children with dilated cardiomyopathy: preliminary report of first two cases. Pediatr Transplant. 2010;14:257-60.

9. Infective Endocarditis

A 9 year old girl is known to have rheumatic heart disease, she presented to the emergency room with fever for 10 days associated with shortness of breath.

P/E

Ill, temp 39 degrees, RR 40 /min, HR 120 /min. Throat: clear
Chest: clear
CVS; Signs of heart failure, Pansystolic apical murmur with mid diastolic soft murmur,
Liver: 5 cm below the costal margin with a span of 10 cm, spleen 3 cm below the costal margin.

Investigations:

CBC: TWBC 19000/hpf , Neutrophilia
ESR 80 mm/Hour. Urine: RBC 10-15 /hpf
ASO: Less than 200
Blood cultures: 3 samples were taken.
Echo: Severe mitral regurgitation. A vegetation was seen on the anterior mitral valve leaflet measuring 4x6 mm.
Diagnosis:
Definite Infective endocardities (IE) (one major and 2 minor criteria)
NB: Recurrence of ARF is ruled out by a low ASO titre.

Discussion:

The epidemiology of IE had changed a lot in the last century and this is attributed to the increased survival of patients with operated and palliated heart diseases. On the other hand the number of patients who are subjected to invasive procedures and central lines has increased dramatically which resulted in increasing number of patients with procedure-based IE. The most common organisms are streptococcus viridans followed by staphylococcus. It is very important to document the diagnosis of IE as the treatment is time and resource consuming.

The Modified Duke's Criteria:

Major criteria:

1. Histopathological evidence of IE from excised heart valves
2. Positive blood culture with an organism consistent with BE e.g strep viridians
3. Echocardiographic evidence of vegetations (valvular or mural), periannular abscess or new dehiscence of prosthetic valves.

Minor Criteria:

1. Intermittent bacteremia or fungemia.
2. Fever.
3. Major embolic events. (major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages,and Janeway's lesions)
4. Nonembolic vascular phenomena(glomerulonephritis, Osler's nodes, Roth's spots)
5. Underlying valvular disease, injection drug use or indwelling catheters.
6. Echocardiographic abnormalities that fell short of typical lesions described above.

Criteria for diagnosis:

Definite IE:

2 major criteria; or
1 major criterion and 2 minor criteria; or
5 minor criteria

Possible IE

1 major criterion and 1 minor criterion; or
3 minor criteria

Rejected (one of the following)

Firm alternative diagnosis explaining the features of IE.
Resolution of IE syndrome with antibiotic therapy for 4 days or less.
No pathological evidence of IE at surgery or autopsy, with antibiotic
therapy for 4 days or less; or Does not meet criteria for possible IE as above.

Management:

- Supportive: analgesics, antipyretics and management of CHF.
- Antibiotics: first choice of antibiotics is penicillin plus gentamicin for 2 weeks then penicillin for another 4 weeks. In patients with recent history of cardiac surgery especially with prosthetic material vancomycin should be used. Duration of antibiotics: 4-6 weeks guided by organism sensitivity and patient response.

Indications for surgical intervention: patients with large vegetations (>10 mm) and those who failed to respond to medical treatment (nonresolving fever or heart failure) should be referred for surgery.

Prevention:

- IE prevention begins with good dental hygiene and IE prophylaxis in the form of one dose of antibiotic taken 1 hour prior to procedures that provoke bactremia (e.g. dental examination).
- Some recent recommendations underestimated the role of IE prophylaxis but these reports were met with criticism as IE is a serious disease which should be prevented by all possible methods.

High-Risk Category:

Prosthetic cardiac valves, including bio-prosthesis and homograft valves

Previous bacterial endocarditis

Complex cyanotic congenital heart defect (e.g. TGA, TOF)

Systemic-to-pulmonary artery shunt or conduit

Moderate-Risk Category

Most other congenital heart defects (e.g., PDA, VSD,AVSD, CoA, bicuspid aortic valve)

Acquired valvular dysfunction (e.g., rheumatic heart disease, collagen vascular disease)

Hypertrophic cardiomyopathy

Mitral valve prolapse with MR and/or thickened mitral valve leaflets

Negligible-Risk Category (prophylaxis not indicated)

Isolated secundum ASD

Surgical repair of ASD,VSD and PDA (without residua beyond 6 months)

Previous coronary artery bypass surgery

Mitral valve prolapse without mitral regurgitation

Innocent heart murmurs

Previous Kawasaki disease without valvular dysfunction

Previous rheumatic fever without valvular dysfunction

Cardiac pacemakers (intravascular and epicardial) and implanted defibrillators

Suggested Readings:

1.de Sa DD, Tleyjeh IM, Anavekar NS, Schultz JC et al.Epidemiological trends of infective endocarditis: a population-based study in Olmsted County, Minnesota. Mayo Clin Proc. 2010;85:422-6.

2.Baddour LM et al.Infective endocarditis: diagnosis, antimicrobial therapy, and management of complications: a statement for healthcare professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, and the Councils on Clinical Cardiology, Stroke, and Cardiovascular Surgery and Anesthesia, American Heart Association: endorsed by the Infectious Diseases Society of America. Circulation. 2005 14;111:e394-434.

10. Kawasaki Disease

A 10 months old boy presented with fever for 10 days, he received oral antibiotics with no response.

P/E

Ill, irritable, Temp 39 degrees, RR 40/min, HR 130/min. Generalized polymorphous , maculopapular skin rash , red eyes , red oral cavity with lip fissuring. Cervical lymphadenopathy.

Investigations:

Total white cell count 18000/ml3, ESR 90mm /hour, Platelet 800000/ml3.

Urine: uncountable pus cells , Culture : no growth

Echo: Dilated left main coronary artery measuring 5 mm (Fig 64). Normal right coronary artery. Mild mitral regurgitation. Small pericardial effusion.

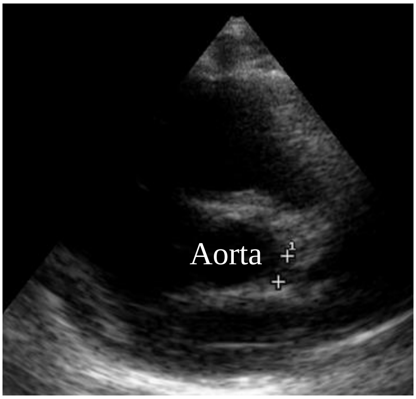


Figure 64

Echocardiographic short axis view of the Ao showing dilated left main coronary artery measuring 5 mm (normal up to 3 mm)

Diagnosis:

Kawasaki disease.

Management:

- Intravenous Immunoglobulins:2g/kg/dose over 12 hours
- Aspirin: 30-50 mg/kg/d divided 6 hourly till the acute phase is over (ESR comes down) then reduce to 5 mg/kg/day till coronary dilatation improves.

Discussion:

Diagnostic Criteria:

Fever for more than 5 days accompanied by four out of five of the following signs:

1. Red eyes, bilateral non-purulent conjunctivitis.
2. Sore erythematous oral cavity, red fissured lips and classically a‘‘strawberry’’ tongue with protuberant papillae.
3. Polymorphous maculopapular skin rash.
4. Cervical lymphadenopathy, usually unilateral and .1.5 cm in diameter.
5. Erythema and oedema of the hands and feet. Desquamation of the skin is seen to occur during the later stage of the illness.

Other Common Features:

Irritability, possibly secondary to cerebral vasculitis or to the presence of aseptic meningitis.

Mood alterations, behavioral changes.

Gastrointestinal upset and diarrhea (20%).

Pain or arthritis affecting large joints (30%).

Reported Sequelae:

Hydrops of the gall bladder (10%)

Deranged liver function

Heart block

Myocardial dysfunction.

Differential Diagnoses
Toxic shock syndrome
Staphylococcal scalded skin syndrome
Scarlet fever.
Viral exanthemas.
Juvenile chronic arthritis.
Infection with adenovirus, Coronavirus, measles, parvovirus and rickettsiae.

Incomplete KD:

- Patients satisfying only 3 out of 5 criteria but at a high risk of coronary involvement
- Usually no cervical lymphadenopathy
- Common in < 1 year and > 4 year of age.

Management:

- Aims of management:

1. To reduce arteritis.
2. To inhibit platelet aggregation thus prevent arterial thrombosis.

- IVIG (as above)
- Nonresponders: repeat IVIG 2 g/kg/dose, cytotoxic drug had been used (methotrexate, cyclosporine)
- Aspirin (as above)This is continued for 6 weeks if there is no coronary involvement and longer if coronary involvement persists
- Anti cytokine drugs: infliximab had shown benefit in refractory cases.
- Follow up by echo.

- If giant coronary aneurysms are present treatment with anticoagulation drugs should be started with close follow up.
- Long term follow up of children with KD without coronary aneurysms had shown favorable results. Patients with giant coronary aneurysms has shown lower physical activity scores compared to normal children.

Suggested Readings:

1.L E Wood and R M R Tulloh. Kawasaki disease in children. Heart 2009;95:787–792.

2.Baker AL, Gauvreau K, Newburger JW, Sundel RP et al. Physical and psychosocial health in children who have had Kawasaki disease. Pediatrics. 2003;111:579-83.

11.Pericarditis

A 6 year old girl came to the emergency room with acute respiratory distress. There was history of fever and runny nose for the last 10 days, then she developed chest pain and shortness of breath which became progressive.

P/E

Ill, distressed, respiratory rate 40/min, Heart rate: 130/min with week peripheral pulses. BP: 70/60 (narrow pulse pressure).

Temp: 38 degrees.

CVS: Distant (muffled) heart sounds, no murmurs, a friction rub is heard. Raised jugular venous pressure. Liver: 6 cm below the right costal margin, span 12 cm.

CXR: Massive cardiomegaly

ECG: Low voltage

CBC: Leukocytosis with dominant lymphocytes

ESR: 60 mm/hour

Echo: Figure 65

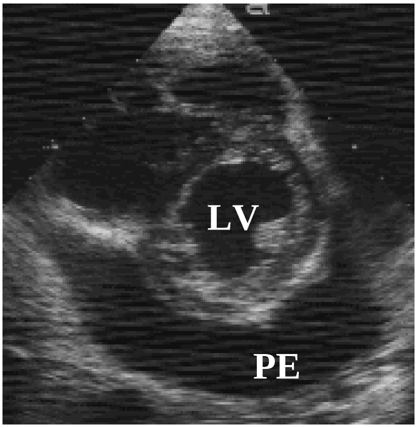


Figure 65:

Echocardiographic short axis view showing a large pericardial effusion (PE) with clear fluid.

The patient underwent pericardiocentesis under echo and noninvasive monitoring. 300 ml of clear straw-colour fluid was drained.

Fluid analysis:

Protein: 3 gm/dl

Cell count: 150/hpf, lymphocytes were 80%

Fluid gram stain and culture: Negative.

Diagnosis: Viral pericarditis

Management:

Confirmation: viral antibody titres: the most common is Coxsackie viruses, other viruses include echovirus, adenovirus, Epstein-Barr virus, influenza virus, and human immunodeficiency virus.

The patient was put on antibiotics (ampiclox plus ceftriaxone) till all cultures (blood and pericardial fluid) came negative. Non steroidal anti inflammatory drugs were given for pain relief. Follow up echo at 5 and 15 days showed no pericardial effusion.

Discussion:

The differential diagnosis:

1. Bacterial pericarditis: rarer than viral pericarditis, the fever and constitutional symptoms are more prominent. The fluid is turbulent with polymophonuclear leukocytosis and culture may show *S. aureus*, *Streptococcus pneumonia* or *Haemophilus influenzae*. Echo can show a thick, tissue-like fluid with fibrinous strands (Figure 66). This type of effusion needs open surgical window to drain it.

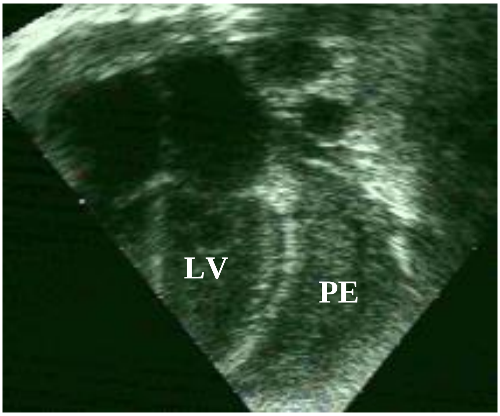


Figure 66:
Echocardiographic 4-chamber view showing a large thick pericardial effusion (PE) that needs to be drained by a pericardial window.

2. Tuberculous pericarditis: This is common in areas where tuberculosis is endemic. It is usually chronic and the patient can present with signs of right heart failure, typically without manifestations of systemic tuberculosis. It can be associated with low grade fever and respiratory distress. The effusion is commonly bloody with high protein and high cell count. Confirmation by pericardial biopsy and polymerase chain reaction test for the blood or pericardial fluid is needed. If suspicion is high a trial of anti tuberculosis drugs should be given. Treatment with anti tuberculosis drugs plus a 6-weeks course of steroids is indicated. The most serious complication is tuberculous constrictive pericarditis which is due to pericardial fibrosis and calcification. It is managed by surgical resection of the pericardium. (pericardiectomy)
3. Acute rheumatic fever: can present with pericarditis , the pericardial effusion is usually small-moderate and rarely

needs drainage. Pericarditis is almost always associated with valvular lesions.

4. Connective tissue diseases: a relatively common cause of pericarditis.
5. Severe CHF: right sided failure and other causes of severe HF e.g. in cardiomyopathy can be associated with pericardial effusion.
6. Other causes: renal failure, malignancies especially lymphoma, post-radiation, hypoalbuminemia, post cardiac surgery (post pericardiotomy syndrome) and drug induced (hydralazine, isoniazid, and procainamide).

Recurrent Pericarditis:

In 15% of patients pericaditis can be recurrent and chronic. Many drugs have been tried like nonsteroidal anti-inflammatory drugs and steroids. Colchicine have been tried for chronic pericardidits with favourable results.

Further Readings:

1.Lange RA, Hills DL. Acute pericarditis. *N Engl J Med*. 2004;351:2195–2202.

2. Blanco CC, Parekh JB. Pericarditis. Pediatr Rev. 2010;31:83-4

12. Supraventricular tachycardia

Scenario 1

SVT in infants:

A 4 weeks old infant was brought to the emergency room because the mother noticed irritability, poor feeding and frontal sweating.

P/E

Irritable, Temp 37 degrees, HR is very fast, BP 80/60. The patient was connected to the cardiac monitor: the heart rate was found to be 300 beats/minute, ECG was done (figure 67)

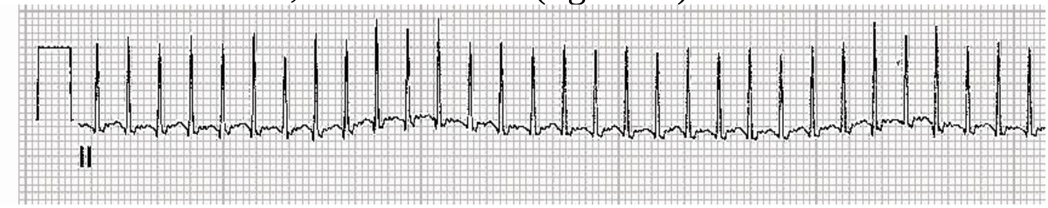


Figure 67

Rhythm strip of the patient showing a narrow, regular QRS, heart rate 300 beats/min, P wave is seen at the end of the QRS.

Diagnosis:

Atrioventricular re entry tachycardia (AVRT)

Features:

1. Narrow QRS, P wave is not seen before but at the end of the QRS.
2. Very regular heart rate with no change with breathing or temperature.
3. Rapid onset and rapid termination.
4. Response to vagal maneuvers and adenosine.

Management:

The patient is hemodyamically stable so:

- Keep connected to ECG.
- Vagal maneuvers: bring a large bowel with ice-cold water and hold the baby tightly with the face down and immerse the face briefly in the bowel.
- Can put a bag with ice cold water on the forehead
- Unilateral carotid sinus message
- Avoid pressure on the eyeball.
- Give Adenosine by rapid IV bolus followed by a saline flush. Use a 3 way valve and prepare the adenosine and the flush, one person administers the adenosine and the other person gives the saline immediately after. The dose is 50 μg/kg, increments of 50 μg/kg are given every 1 to 2 minutes. The usual effective dose is 100 to 150 μg/kg with maximum dose of 250 μg/kg.
- If the BP is low: stabilize the patient (ABC), emergency immediate cardioversion by 0.5 joule/kg increasing in steps up to 2 joule/kg.

Alternative drugs in stable patients:

- Digoxin can be used in infants *without* WPW
- In the presence of WPW intravenous esmolol or propranolol is used, the antidote for propranolol is isoprenaline which should be given if there is severe bradycardia or hypotension.

- IV verapamil **should be avoided** in infants and young children because it may produce extreme bradycardia and hypotension leading to mortality.
- Digoxin should be avoided in patients with WPW

Prevention:

- If the resting ECG is showing WPW: use betablockers (e.g. propranolol) or flecainide.
- If WPW is not present digoxin can be used in young infants.
- Avoid digoxin and verapamil in WPW as it may (by blocking the AV node) increase the rate of antegrade conduction of the impulse through the accessory pathway leading to ventricular fibrillation, however this is extremely rare in the paediatric age.

Scenario 2

A child with palpitations

A 12 year old girl came complaining of frequent episodes of palpitations, they come and go abruptly and cause her to feel unwell, dizzy and mildly sweaty but there is no loss of consciousness.

P/E

Normal vital signs, HR 80/min, well looking, normal growth.

CVS: normal S1 and S2, no murmurs. Not in CHF

No tremors or thyroid enlargement.

ECG: Figure 68

Echo : normal (needs to rule out Ebstein;s anomaly, congenitally corrected TGA and mitral vavle prolapse)

Holter monitor: normal.

CBC and thyroid function: normal

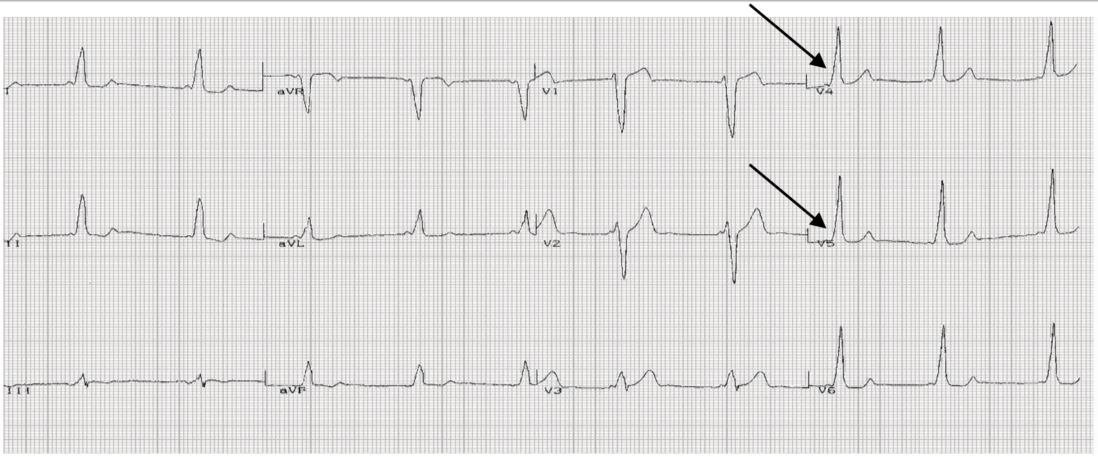


Figure 68:
ECG showing WPW: a short PR interval (0.06 seconds) and a delta wave best seen in V4,5 and 6 (arrows)

The patient was and asked to try and record the ECG during the episode of palpitations. Few days later she presented with her ECG during the episode which revealed a HR 170 /min, narrow complex, regular, P wave is abnormal preceding the QRS.
Diagnosis:
AV node reentry tachycardia

Management:
Teach the child vagal maneuvers e.g. the valsalva maneuver.
One of these drugs: Flecainide , propranolol or atenolol.
Electrophysiological study is done to map the arrhythmia and ablate the accessory pathway.

Discussion:

- SVT due to re entry (accessory pathway) is the commonest arrhythmia in the paediatric age. It has 2 peaks: one in the neonate and another in older children. The commonest type is atrioventricular re entry tachycardia (60-70%) followed by atrioventricular *node* re entry which is more common in older children. SVT rarely leads to life threatening complications and the most common symptoms in infants is irritability while older children often complain of palpitations.

- Documentation of SVT is very important, 12 lead ECG, Holter and exercise ECG can delineate the arrhythmia, however if episodes are infrequent it can be difficult to document.
- Event monitor (when available) is a very useful device that the patient takes home and when symptoms occur the patient starts recording then transmits the ECG by telemetry to the treating hospital .
- Implantable loop recorders are small recording devises that can be implanted subcutaneously to record complex serious arrhythmias (Fig 17)
- In newborn babies and infants, pharmacological therapy is advised due to the high spontaneous cessation rate of those tachycardias at the end of the first year of life. In contrast, the probability of spontaneous cessation of tachycardia in children > 1 year of age is very low. Therefore, radiofrequency catheter ablation of the anatomical substrate of the tachycardia is a rational alternative to long-lasting antiarrhythmic therapy. Results in children with a structurally normal heart are comparable to those achieved in adults.
- Medical treatment of SVT in paediatric age is still evolving as large scale trials are lacking. Most data are derived from adult trials. Beta blockers and falecainide are the first line drugs. Digoxin and verapamil **should be avoided** in patients with WPW. If there is no evidence of WPW digoxin can be used especially in infants and

verapamil in older children. Care to proarrhthmic side effects like prolonged QTc interval should be paid.

Rarer Causes of SVT:

- Atrial flutter (Figure 69) very rare except in post operative patients. The most important findings clinically is irregular heart rate in contrast to very regular re entry SVT.

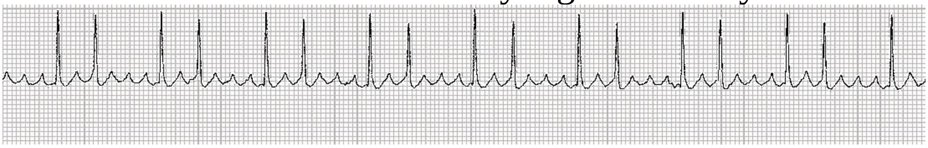


Figure 69:
Rhythm strip showing atrial flutter: atrial contractions (saw-tooth appearance) with a rate of 300/min with irregular QRS.

- Atrial fibrillation: rare in infants. Common in children with severe mitral valve disease (Figure 55)

Suggested Readings:

1.Paul T, Bertram H, Kriebel T, Windhagen-Mahnert B et al. Supraventricular tachycardia in infants, children and adolescents: diagnosis, drug and interventional therapy. Z Kardiol. 2000 ;89:546-58.

2.Lee PC, Hwang B, Chen SA, Tai CG et al. The results of radiofrequency catheter ablation of supraventricular tachycardia in children. Pacing Clin Electrophysiol. 2007 ;30:655-61.

3.B.J. Deal ,C.L. Johnsrude ,S.H. Buck.Pediatric ECG Interpretation :An Illustrative Guide. Blackwell, Futura . First Ed, 2004.

4.Sulafa KM Ali Collateral effects of anti arrhythmic drugs in children. Cur Pharmac Design; 2008;14:782-787.

13. Bradycardia

You were called to a neonate just born with slow heart rate. Pregnancy was uneventful; the mother has no symptoms suggestive of systemic lupus (SLE) however her previous baby suffered from shortness of breathing and was diagnosed as ‘cardiomyopathy’. He died at the age of 3 months. The obstetrician detected fetal bradycardia at 36 weeks therefore the baby was delivered by emergency caesarean section .

P/E

The baby looks well, the birth weight is 2.5 kg. Heart rate is 50/min, normal BP, O2 saturation 99%., normal S1and S2, no murmur. No signs of CHF.

ECG: (Fig 70)

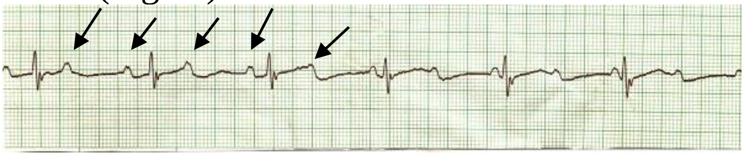


Figure 70:
ECG rhythm strip showing complete AV block: the P wave (arrows) is completely dissociated from the QRS, P wave rate is 100/min and QRS rate 50/min.

Echo: normal anatomy. The ejection fraction was low (40%). Few weeks later the baby developed papular erythematous skin lesions distributed in the face and trunk. The skin lesions faded spontaneously after 2 weeks.

CBC: Normal, no thrombocytopenia. LFT: normal

Further investigations:

- Mother and patient's anti Ro and anti La antibodies were highly positive.

Diagnosis: Neonatal lupus syndrome (NLS)

Management: If the patient presented with severe bradycardia with hypotention a temporary pacing wire is positioned urgently through internal jugular or femoral vein till a perminant pacemaker is implanted epicarial in small infants and transvenous in older patients.

. The patient was put on diuretics as he developed CHF and EF was 30%. He died at home at the age of 4 months.

Discussion:

NLS is a common manifestation of maternal SLE. The most constant manifestation is congenital complete AVB which occurs in 10% of pregnancies of SLE affected mothers. Other manifestations include skin rash, throbocytopenia and liver dysfunction which are usually transient. Early detection of fetal heart block is crucial , it can be achieved by careful ultrasound/echo scanning. Heart block usually start as first degree and progresses, if detected early the mother can be treated by corticosteroids or intravenous immunoglobulins which may help to reverse the fetal heart block. Cardiomyopathy is a rare reported association of NLS that can occur late, some cases show enocardial fibroelastosis.

Other causes of heart block in infants include some CHD like congenitally corrected TGA and isomerism. Complete AV block can follow surgical closure of VSD especially the inlet type. The reported frequency of post operative AVB is 1-3 % in large surgical centres. The patients are managed by temporary pacing through epicardial pacing leads inserted by the surgeon at the time of operation.

Second degree AV block: can occur in complex CHD or following myocarditis or cardiac surgery. MobitzType 1: Wenckibach phenomenon: progressive prolongation of PR interval then a dropped beat, Type II: sudden drop of QRS (more serious). (Figure71)

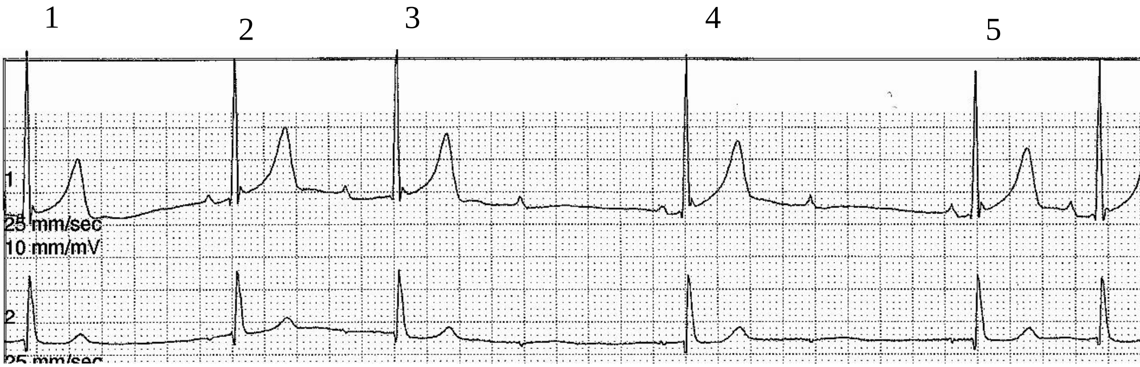


Figure 71
ECG rhythm strip showing two types of second degree AV block: progressive prolongation of PR interval in the second and third beats then a dropped beat (Mobitz II). After the 4th beat there is sudden drop (Mobitz I)

The indications for permanent pacing in complete AVB :

1. All symptomatic patients. Symptoms included heart failure, dizziness and syncope.
2. Heart rate 55 beats/minute or less in asymptomatic patients
3. Post operative complete AVB is managed by permanent pacing if it did not resolve within 10 days of surgery. There is a small risk of late heart block therefore the patients who recovered from transient block should be followed up periodically by clinical and ECG examinations.

Other indications:

Mobitz type II
Sick sinus syndrome
Some cases of Long QTc syndrome.

Other causes of bradycardia:

Sinus bradycardia: can be physiologic during sleep and in athletic adolescents. Pathological causes include hypothyroidism and increased intracranial pressure.

Living with a permanent pacemaker:

- Regular follow up for PM checking and programming
- The battery needs to be changed after certain time depending on the PM check.
- Patients should avoid security detectors at airports and government buildings, MRI and other strong magnetic fields.

Suggested Readings:

1.Krishnan AN, Sable CA, Donofrio MT.Spectrum of fetal echocardiographic findings in fetuses of women with clinical or serologic evidence of systemic lupus erythematosus. J Matern Fetal Neonatal Med. 2008 ;21:776-82.

2.Brucato A, Cimaz R, Caporali R, Ramoni V, Buyon J. Pregnancy Outcomes in Patients with Autoimmune Diseases and Anti-Ro/SSA Antibodies.Clin Rev Allergy Immunol. 2009 [Medline]

3.Buyon JP, Clancy RM. Neonatal lupus syndromes. Curr Opin Rheumatol. 2003 ;15:535-41.

4.Gil J. Gross, MD,Christine C. Chiu, MSc,Robert M. Hamilton, MD,Joel A. Kirsh,Elizabeth A. Stephenson.Natural history of postoperative heart block in congenital heart disease: Implications for pacing intervention.Heart Rhythm 2006;3:601– 604

14. A child with syncope

Scenario 1

A 15 year old boy presented with history of dizziness on exertion, he feels palpitations and sweating followed by loss of consciousness. His older brother died 2 years ago at 18 years of age while playing football.

P/E

Well, HR 60/min, BP 110/70

Normal heart sounds, no murmur. Normal other systems

ECG: Fig (72)

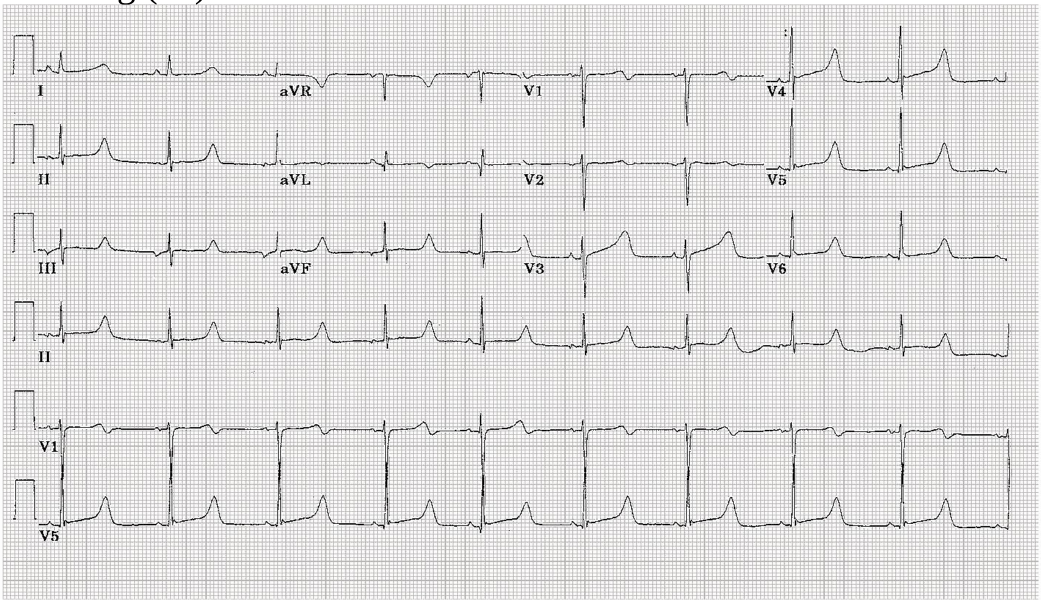


Figure 72:

ECG showing normal sinus rhythm. The QT is 0.56 sec (heart rate 60/min no need for correction of QT)

Diagnosis: Long QT Syndrome (high probability)

Scenario 2

The 10 year old brother of the patient in Scenario 1 presented to the ER with syncope: his BP is irrecordable and peripheral pulses not palpable, ECG monitor showed this rhythm: (Figure 73)

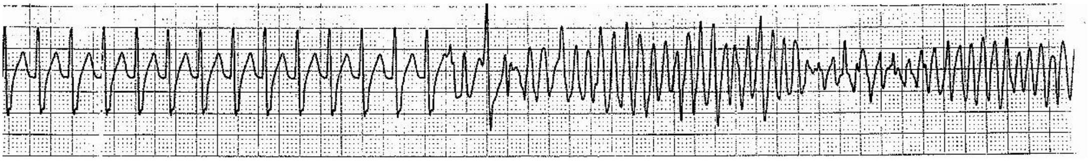


Figure 73:
Rhythm strip showing tachycardia followed by ventricular fibrillation with changing morphology (Torsade de points)

Diagnosis: Ventricular fibrillation –Torsade de pointes
Management:
ABC
CPR, Defibrillation 2,4 then 6 joules /kg were given
Magnesium sulfate 25 to 50 mg/kg, IV was given for Torsade de pointes. The patient converted to sinus rhythm. He was started on propranolol and advised to restrict all forms of competitive exercise. He was referred for cardiovertor defibrillator (ICD) implantation.Family members were called for screening.

Discussion:

- Syncope, defined as sudden transient loss of consciousness,

followed by prompt, complete recovery, is frequently encountered as a presenting problem in children and young adults.

- Cardiac causes include arrhythmias, most important of which are ventricular tachycardia/fibrillation related to long QT and similar syndromes. Other cardiac causes are left ventricle outflow tract obstruction like AS and HOCM.
- More common are non cardiac causes that include vasovagal syncope and epilepsy.

- LQT syndrome is a serious arrhythmogenic disorder that can lead to sudden cardiac death which is potentially preventable
- The presenting complains can be the typical history of syncope with positive family history however some patients may have complains like ‘funny turns’ or convulsions therefore ECG should be carefully examined.
- Diagnostic Criteria for Long QT syndrome

(in the Absence of Medications or Disorders Known to Prolong the QTc Interval)

Diagnosis:

1 point = low probability of LQTS
2 to 3 points = intermediate probability of LQTS
4 or more points = high probability of LQTS

- The ECG should be carefully examined for the QTc as well as for T wave alterations like T wave alternans and notching.
- In some patients standard ECG can be *normal* and they may need more studies (Holter, exercise ECG, epinephrine provocation test and genetic studies) to make the diagnosis.
- Patients diagnosed with Long QT should have family screening ECG/other tests.
- Beta blockers (propranolol) should be started for the commonest type of LQTS , it is effective in 70% of patients. Some patients require addition of other anti arrhythmic drugs like mexiletine. Certain types of LQTS (LQTS3) do not respond to betablockers

Table 13
Criteria for diagnosis of Long QTc syndrome

Criteria	Score
ECG:	
QTc >480 msec	3
QTc 460–470 msec	2
QTc 460–470 msec	2
QTc 450 (male) msec	1
Torsades de pointes	2
T-wave alternans	1
Notched T waves in three leads	1
Low heart rate for age (<2nd percentile)	1
Clinical History	
Syncope	
With stress	2
Without stress	1
Congenital deafness	0.5
Family History	
Family members with definite LQTS	1
Unexplained sudden cardiac death <30 yr among immediate family members	0.5

- Implantable cardioverter- defibrillators (ICD) can prevent sudden death by detection and treatment of ventricular fibrillation. Indicated in high risk patients defined as those with aborted cardiac arrests (like the patient in scenario 2) recurrent cardiac events despite conventional therapy (with beta blockers) and those with extremely prolonged QTc intervals (e.g.>0.60 second).

- Recent genetic studies have shown that certain mutations in sodium and potassium channels are responsible for the common types of LQTS. So far 12 mutations had been identified. There is strong evidence that *SCN5A (defect in sodium channel)* is associated with sudden infant death syndrome. There is a clear genotype-phenotype correlation regarding the triggering factors for ventricular arrhythmia, in LQT1 the triggering factor is usually exercise while in LQT2 the event is provoked by postpartum stress and auditory stimuli.
- Drug induced LQTS should always be considered in patients with long QTc. A large number of drugs is reported to cause LQTS .Common drugs include antiarrhythmics (e.g sotolol and procainamide) some antibiotics like macrolides.The offending drug should be immediately withdrawn.

Other Arrhythmogenic Conditions:

Brugada syndrome: a rare cause of sudden death due to ventricular arrhythmias, seen mostly in Southeast Asian men. The ECG typically shows right bundle branch block with J-point elevation and concave ST elevation in V1 (Figure 74). Implantable ICD is indicated

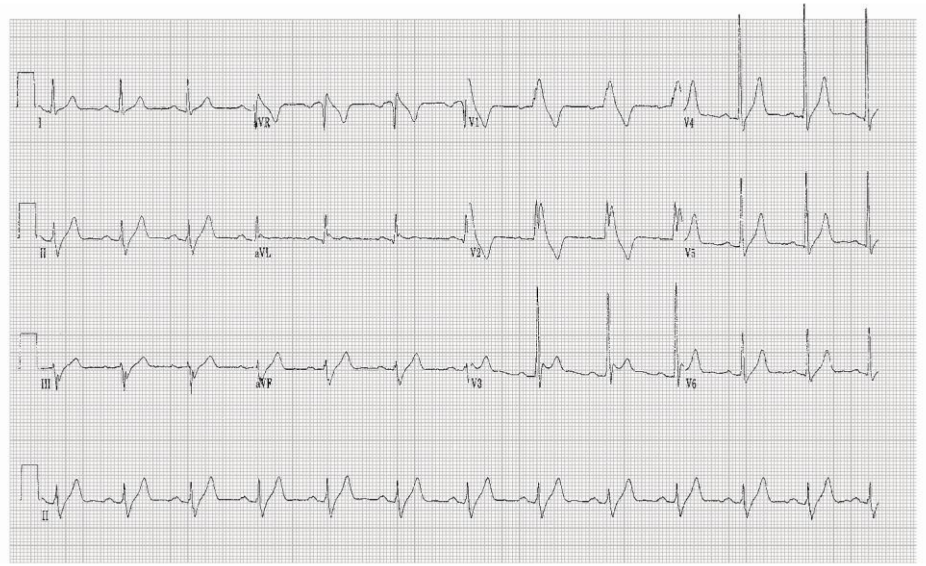


Figure74: ECG from a young adult with Brugada syndrome. Note the right bundle branch block (V1 and 2) and ST elevation in V1.

Short QTc syndrome: is a relatively recently described disorder characterized by a very short QT interval and by susceptibility to atrial and ventricular fibrillation. ICD implantation is the primary therapy.

Non- cardiac syncope

Orthostatic Intolerance syndrome:

An important and relatively common cause of syncope is the Orthostatic Intolerance syndrome which has 3 main types: vasovagal syncope, orthostatic hypotension, and postural orthostatic tachycardia syndrome (POTS). The main symptoms are hypotension /syncope with changing posture, usually upon standing.The Head up tilt table test (Fig 75) is used to diagnose these conditions. Patients lie supine on an electric tilt table and have an intravenous line established. ECG and BP measurements are performed. Patients are tilted into the 60- to 80-degree head-up position for a period of up to 30 minutes with recording of BP every 1 or 2 minutes and continuous heart rate and rhythm monitoring. If a patient becomes symptomatic or the 30-minute time elapses, the tilt table is immediately returned to the supine position.

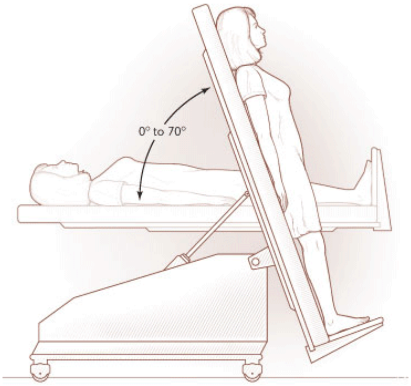


Figure 75:
The head up tilt table test. (Ref no.6)

Types of response:
1. Vasovagal: an abrupt decrease in BP, usually with bradycardia
2. Dysautonomia (or postural hypotension): a gradual decrease in BP leading to syncope
3. POTS: an excessive heart rate increase to maintain an adequate BP to prevent syncope
Treatment of orthostatic Intolerance syndromes: increased salt and water intake. Drugs like fludro-cortisone, alpha blockers, betablockers (meoprolol) or pseudoephedrine can be tried. Permanent pacing is used in patients with refractory and serious symptoms.

Suggested Readings:

1.Schwartz PJ, Moss AJ, Vincent GM, Crampton RS: Diagnostic criteria for the long QT syndrome: An update. Circulation1993; 88:782–784.

2.Collins KK, van Hare GF: Advances in congenital long QT syndrome. Curr Opin Pediatr2006;18:497–502.

3. Patel U, Pavri BB. Short QT syndrome: a review. Cardiol Rev. 2009;17:300-3.

4.Younoszai AK, Franklin WH, Chan DP, Cassidy SC, Allen HD. Arch Pediatr Adolesc Med. Oral fluid therapy. A promising treatment for vasodepressor syncope.1998 ;152:165-8.

5.Deal BJ, Strieper M, Scagliotti D, Hulse E, Auld D, Campbell R, Strasburger JF, Benson DW Jr. The medical therapy of cardioinhibitory syncope in pediatric patients. Pacing Clin Electrophysiol. 1997 ;20:1759-61.

6.Freeman R. Clinical practice. Neurogenic orthostatic hypotension.

N Engl J Med. 2008;358:615-24.

15. Involvement of the Heart in Connective Tissue Diseases

Scenario A

A 5 year old girl presented with painful swelling of the right knee joint for 2 months. For the last week she is complaining of chest pain

P/E

Right knee join is swollen, mild tenderness, restricted movement. No other joint involvement. CVS: Normal S1 and S2, a pansystolic apical murmur is heard radiating to the axilla. A pericardial friction rub is heard.

Investigations:

CBC:TWBC: 15000/hpf, polymorphonuclear cells 60%, Hb: 8gm/dl. ESR: 120/hour. Antistereptolysin O titre, antinuclear antibody and rheumatoid factor: negative.

ECG: Fig 74

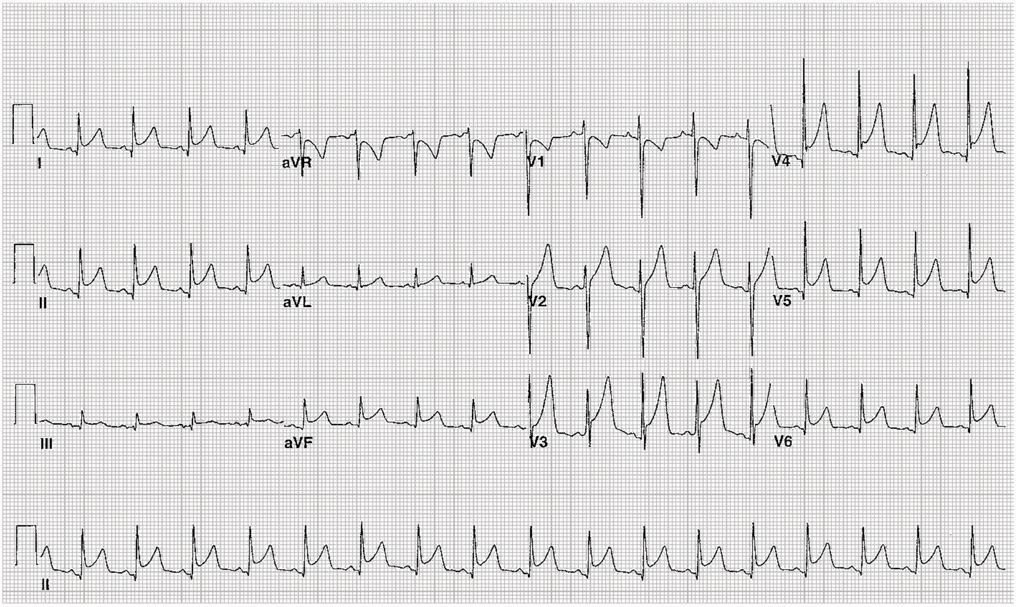


Figure (above) 76:
ECG showing elevated ST segment best seen in leads II,V5and 6

Echo:
Moderate MR. Small pericardial effusion.

Diagnosis:
Pauciarticular juvenile rheumatoid arthritis with MR and pericarditis.
Management :
The patient was started on diuretics, ACE inhibitors and nonsteroidal anti inflammatory drugs. She was referred to the rheumatologist who started her on methotrexate.

Scenario 2
A 12 months old infant presented with fever for 3 weeks associated with skin rash and irritability.
P/E
Irritable, Temp 39 degrees, RR 50/min, HR 150/min
Maculopapular erythematous skin rash all over the body and extremities. The right knee joint is swollen and tender. .
Abdomen: Hepatosplemomegaly

Investigations:
WBC: 18000 with polymorph predominance,ESR 130 mm/h,
Echo: small pericardial effusion
Diagnosis:
Systemic onset juvenile chronic arthritis with pericarditis.
Differential Diagnosis: Kawasaki disease

Discussion:
Connective tissue disease (CTD) can be associated with a range of cardiac abnormalities. Juvenile Idiopathic Arthritis (JIA) had

been associated with AR and MR which can be severe enough to necessitate valve surgery. Systemic Lupus Erythmatosus (SLE) is associated with pericarditis, valve regurgitation and pulmonary hypertension. Sterile vegetations (Libman Sack’s endocarditis) can be found on echo and should be differentiated from IE. (Figure 77) Myocarditis and cardiomyopathy are known associations of CTD. .Patients with CTD should be routinely evaluated by the cardiologist and echo should be performed. On the other hand cardiac manifestations can be the first sign of CTD therefore CTD should be considered in patients with un explainable valve regurgitation or pulmonary hypertension. Differentiation from the more common acute rheumatic fever can be made from the history which reveals short history migratory arthritis of large joints in acute rheumatic fever while in JCA and SLE the course of the disease is prolonged with involvement of the small joints.

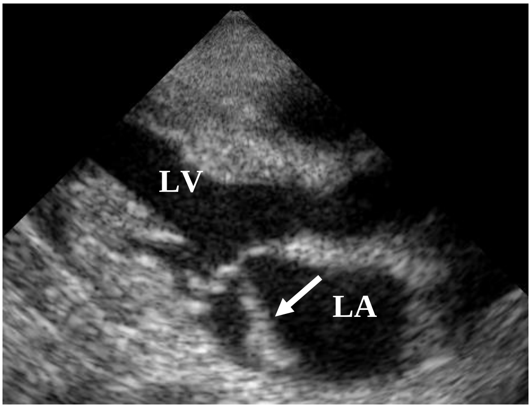


Fig 77
A long axis echo view of a patient with SLE showing a long mass attached to the anterior mitral leaflet (arrow)(Libman Sacks endocarditis)
LA: Left atrium, LV: Left ventricle.

Suggested Readings:

1.Bakkaloglu A, Söylemezoğlu OOzme S, , Alehan DOzer S

Mitral and aortic insufficiency in polyarticular juvenile rheumatoid arthritis. Pediatr Cardiol. 1994;15:151-3.

2. Kramer PH, Imboden JB Jr, Waldman FM, Turley K, Ports TA. Severe aortic insufficiency in juvenile chronic arthritis. Am J Med. 1983;74:1088-91.

3.Yeh TT, Yang YH, Lin YT, Lu CS, Chiang BL.Cardiopulmonary involvement in pediatric systemic lupus erythematosus: a twenty-year retrospective analysis. Microbiol Immunol Infect. 2007;40:525-31.

4.Sulafa KM Ali, Musa M,Noor, Yassmin M O Taha.Cardiac involvement in Sudanese children with connective tissue disease,Sudan Med J2010;46 (2): 73-78

16. Adults with CHD

Scenario 1

A 30 year old lady presented with shortness of breathing on mild exertion. P/E Normal vital signs, O2 saturation 96%. Normal S1, wide splitting of P2. ESM at the pulmonary area.

ECG: RSr pattern, RV volume overload, (figure 78)

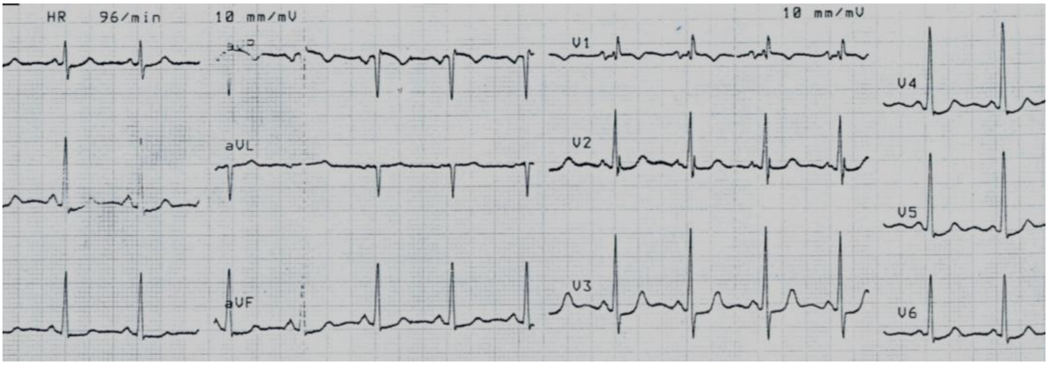


Figure78:
ECG of a patient with ASD showing rsR pattern in V1 (RVH volume overload)

CXR: Cardiomegaly with RV and RA enlargement, plethoric lungs, prominent main PA

Echo: large ASD secundum 25 mm, central defect with good rims.

The patient underwent transesophageal echo in the cath lab then transcatheter closure was done using 30 mm ASD occluder. (Fig79)

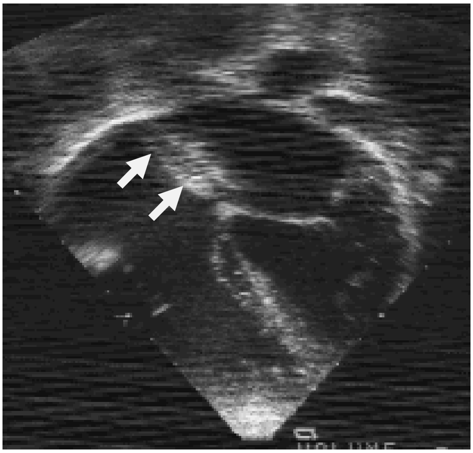


Figure 79:
Echocardiographic 4-chamber view showing the ASD occluder well positioned, aligned with the atrial septum (arrows)

Differential diagnosis:

1. Primum ASD can present at this age. It is a partial form of AVSD and can be associated with left atrioventricular valve regurgitation (due to a pseudo cleft). The ECG is diagnostic with a superior left axis and rsR pattern in V1 (RV volume overload) as in figure 81.

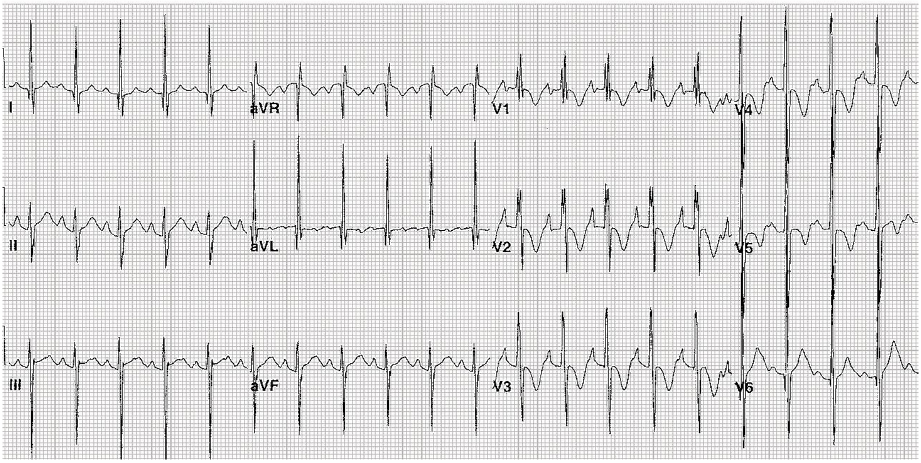


Figure 80 (above) :

ECG of a patient with partial AVSD showing: left superior axis, RVH (volume overload pattern) and LVH which is secondary to left AVV regurgitation.

2.Other types of ASD: (Figure 81):

-Sinus venosus ASD: A defect in the most superior part of the atrial septum at the entry of the superior vena cava associated with partial anomalous PVD (a rare type can occur at the most inferior part of the septum near the inferior vena cava). Clinical features can be similar to secundum ASD except that the O2 saturation is low/normal due to partial APVD. ECG shows a characteristic superior P wave axis.

-Unroofed coronary sinus: a rare type of ASD. Clinical findings are similar to other types of ASD.

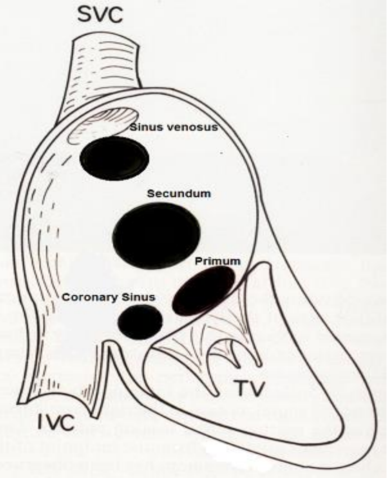


Figure 81:

A diagram showing the positions of the different types of ASD seen from the right atrial side.

3.Pulmonary stenosis: although the site of the murmur of PS is the same as ASD but the murmur of PS is harsher and splitting absent. The ECG shows pressure overload pattern (Figure 51)

Discussion:

ASD is one of the common CHD that presents in adults at the 3rd or 4th decade of life. In children the most common cause of referral is a heart murmur. The time of closure in asymptomatic patients is before school entry and there is no indications to close simple ASD in infancy. In adults the most common symptoms are palpitations and shortness of breathing (CHF). The closure approach depends on echocardiographic anatomy: primum, sinus venosus and coronary sinus defects are closed surgically. Secundum defects are the most common type (75%) and most of them are amenable to transcatheter closure (Figure 79) except those with deficient rims. Follow up studies of large cohorts of patties who had trans catheter closure had shown excellent success rates, however the adverse effects and rate of rescue operations is higher than that of surgical closure. The main complications are device embolization, thrombo-embolism and injury to aortic valve. The device design and implantation techniques are still evolving and gradually improving.

Scenario 2

A 35 year old man had been operated as a child for TOF. He was on regular follow up till the age of 15 years and was asymptomatic. Recently he started to develop palpitations which were associated with dizziness.

P/E

HR 80/min, normal BP. O2 saturation 98%. Normal S1 and S2. To and fro (Ejection systolic with early diastolic) murmurs were heard at the pulmonary area.

ECG: Complete RBBB,sinus rhythm

Echo: Dilated RV with impaired RV function, free pulmonary regurgitation (PR).Peak right ventricle outflow tract gradient is 30mmhg. No residual VSD, normal LV function.

Further Investigations:

- Cardiac magnetic resonance imaging (CMRI): the RV was found to have moderately impaired function, there was severe pulmonary regurgitation. Pulmonary artery branches were normal in size with no peripheral stenosis.
- Holter monitor: no evidence of ventricular arrhythmia.

Diagnosis: RV dysfunction secondary to severe PR

Treatment: The patient was put on antifailure treatment and referred for pulmonary valve replacement.

Discussion:

Surgical repair of TOF started in 1960. Initially, RV incision and PV transanular patch were freely used resulting in free PR. Subsequently, PR was found to have deleterious effects on RV size and function leading to arrhythmias that can cause sudden death. The current trends is to preserve the PV annulus or use a valved patch in order to avoid free PR. Long term follow up had shown that the 35 year survival rates are approaching 85%, however there is a risk of late sudden death of about 2 per 1000 patient-years. Some factors that lead to increased risk of complications post TOF repair are:

1) Long-standing palliative shunts.
2) Older age at the time of definitive surgery.
3) Abnormal RV hemodynamics (due to pulmonary regurgitation and/or residual outflow obstruction)
4) High-grade ectopy on Holter monitor
5) Inducible ventricular tachycardia at electrophysiological study.
6) QRS duration > 180 ms

Life-long follow up is indicated in all patients with repaired TOF. Cardiovascular magnetic resonance (MR) has evolved into an imaging technique that provides adequate and unique, information about the RV size and function. Indications for pulmonary valve implantation are the presence of severe pulmonary regurgitation and any of the following:

1. Moderate to severe RV dysfunction.
2. Moderate to severe RV enlargement.
3. Development of symptomatic or sustained atrial and/or ventricular arrhythmias.
4. Moderate to severe TR.

Transcatheter valve implantation is now becoming increasingly performed with very good results.

Suggested Readings:

1. Di Bardino DJ, McElhinney DB, Kaza AK, Mayer JE Jr.Analysis of the US Food and Drug Administration Manufacturer and User Facility Device Experience database for adverse events involving Amplatzer septal occluder devices and comparison with the Society of Thoracic Surgery congenital cardiac surgery database. J Thorac Cardiovasc Surg. 2009 Jun;137(6):1334-41.

2.Carole A et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart) JACC2008; 52 (23):e8-108.

3.Nollert G, Fischlein T, Bouterwek S, Böhmer C, Klinner W, Reichart B.Long-term survival in patients with repair of Tetralogy of Fallot: 36-year follow-up of 490 survivors of the first year after surgical repair. J Am Coll Cardiol. 1997 Nov 1;30(5):1374-83.

4.He GW.Current strategy of repair of Tetralogy of Fallot in children and adults: emphasis on a new technique to create a monocusp-patch for reconstruction of the right ventricular outflow tract. J Card Surg. 2008 Nov-Dec;23(6):592-9.

5. Lurz P, Gaudin R, Taylor AM, Bonhoeffer P.Percutaneous pulmonary valve implantation. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2009:112-7.

17. A Child with the Metabolic Syndrome

A 12 year old boy was found to have high BP and referred for cardiac evaluation. He is a symptomatic.
P/E
Weight is 70 kg, Height is 158 cm, BMI is 56 (97th % is 32)
BP 130/80
CVS: Normal S1 and S2, no murmurs
ECG: LVH
Echo: Interventricular septum and LV posterior wall thickness 12 mm (normal 8mm).
Fasting blood glucose: Normal
Fasting lipid profile:
Total cholesterol 205 mg/dl (>90%)
LDL cholesterol 130 mg/dl (>90%)
HDL cholesterol 50 mg/dl (50%)
Triglycerides 130 mg/dl (95%)

Diagnosis:
Metabolic syndrome: obesity, hypertension and hyper cholesterolemia**.**

Management:

1. Life style modifications:
    - i. Diet: low caloric, low sodium and low cholesterol
    - ii. Weight reduction
    - iii. Exercise
    - iv. Quit smoking (active and passive)

II.Medications: if above measures did not control cholesterol and BP consider cholesterol lowering drugs. Other causes of hypertension (renal/endocrine) should be ruled out. If BP is still high anti hypertension medications should be started.

Discussion:

Cardiovascular risk factors are conditions that predispose to ischemic heart disease, namely:

Cigarette smoking
Hypercholesterolemia
Hypertension
Low levels of high-density lipoprotein
Diabetes mellitus
Family history of premature ischemic heart disease (before age 55years)

These factors were shown to affect vascular structure and function in early childhood, therefore it is vital to actively control these factors in children. When these risk factors occur in clusters in the same individual, the condition is known as the metabolic syndrome. The combination of risk factors predisposes individuals to the development of coronary artery disease (CAD) and type 2 diabetes. Obesity is a risk factor for CAD independent of the standard risk factors, probably through the emerging risk factors which include atherogenic dyslipidemia (also known as the “lipid triad,” consisting of raised triglycerides and low-density lipoprotein [LDL] particles and low levels of high-density lipoprotein [HDL] cholesterol), insulin resistance (hyperinsulinemia), a proinflammatory state (elevation of serum high-sensitivity C-reactive protein), and a prothrombotic state (increased amount of plasminogen activator inhibitor 1). The cluster of these risk factors occurring in one person is known as the metabolic syndrome.

Definition of Metabolic Syndrome in Adolescents:

1. Triglycerides >110 mg/dL

2. HDL cholesterol <40 mg/dL
3. Waist circumference >90th percentile or body mass index (BMI) >95th percentile
4. Fasting glucose >110 mg/dL
5. Systolic blood pressure >90th percentile for age and gender.

The presence of at least three of the preceding abnormalities constitutes metabolic syndrome This syndrome occurs more commonly in individuals with abdominal (visceral) obesity.

Diagnosis of obesity:

The BMI (weight in kg divided by the square of the height in meters, kg/m2) is a simple, valid measure of relative weight and is recommended in clinical diagnosis of overweight states. The BMI charts for boys and girls reference values are shown in Appendix V.

Drug therapy is indicated in children aged 10 years and older if, after an adequate trial of diet therapy (6 months to 1 year):

1. The LDL level remains at or above 190 mg/dL, with a negative or unobtainable family history of premature coronary CV death.
2. The LDL level remains at or above 160 mg/dL *plus*:
a. Positive family history of premature CVD (before 55 years of age in men and before 65 years in women).
b. Two or more other CVD risk factors (e.g., low HDL levels, cigarette smoking, high blood pressure, obesity, diabetes) or metabolic syndrome is present

The main drugs used are bile acid sequestrants (cholestyramine) and Statins.

Suggested Readings:

1.Berenson GS, Srinivasan SR, Bao W, et al: Association between multiple cardiovascular risk factors and atherosclerosis in children and young adults. N Engl J Med 1998; 338:1650-1656.

2.Cook S, Weizman M, Auinger P, et al: Prevalence of a metabolic syndrome phenotype in adolescents:Findings from the third National Health and Nutrition Examination Survey, 1988–1994. Arch Pediatr Adolesc Med 2003; 157:821-827.

Chapter 4

The Difficult Made Easy

Complex CHD

- Refers to combination of major heart defects.
- Often involves hypoplasia or absence of one ventricle (functionally single ventricle) where the pulmonary and systemic blood supplies are provided by the same chamber.
- The most important point in classifying complex CHD is the presence or absence of ***pulmonary stenosis***. Clinical classification by inspection is very helpful to differentiate the 2 categories therefore plan the management.
    i. If there is PS the lungs are protected and the patient behaves like TOF (cyanosis without CHF)
    ii. If lungs are not protected (mild or no PS) the patient develops heart failure and mild cyanosis.

Examples of complex CHD:

Functionally Single Ventricle:

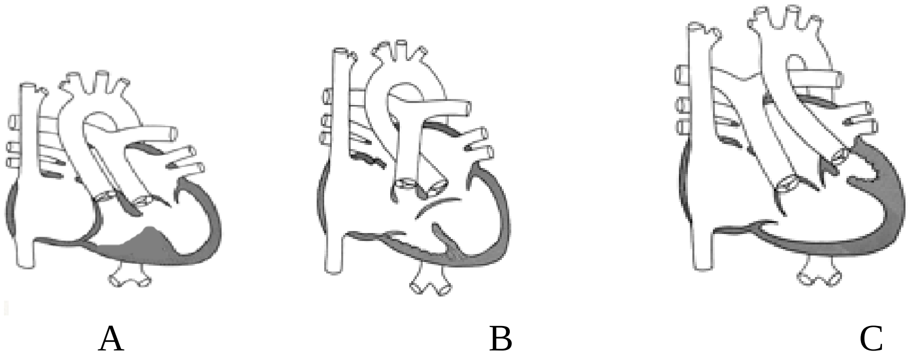


Figure 82
Common types of functionally single ventricle: A: tricuspid atresia
, B: unbalanced AVSD, C: double inlet left ventricle with small RV.

1.Tricuspid atresia:

The tricuspid valve is atretic or hypoplastic with RV hypoplasia (Figure 82-A and 84). There is usually associated VSD and there must be an ASD to allow systemic venous blood to go to the left side. If there is no pulmonary stenosis the left ventricle pumps blood with the same pressure to both aorta and pulmonary artery so severe pulmonary overflow (manifesting as CHF) and cyanosis (due to mixing of venous blood with pink blood in the LV) will ensue.

If there is PS the patient will be TOF-like. Tricuspid atresia is often associated with pulmonary atresia (hypoplastic right heart syndrome). Other associations include TGA (as in Figure 82-A) which can lead to subaortic stenosis as the aorta is arising from the hypoplastic RV.

The ECG is diagnostic (Figure 83)

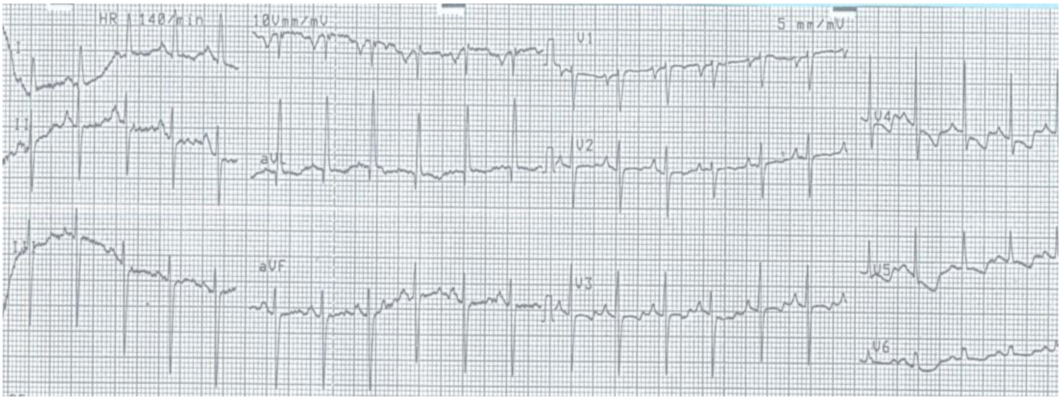


Figure 83:
ECG showing superior left axis(positive voltage in lead I and negative in aVF), right atrial enlargement and small RV forces (negative voltage in lead V1) in a 2 month old with tricuspid atresia.

Management:

Staged palliation (see below)

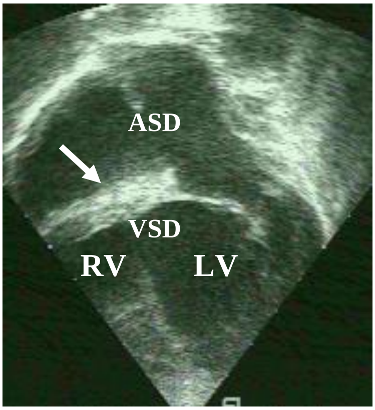


Figure 84:
Echo 4 chamber view showing the atretic TV (arrow), small RV, large ASD and VSD.

2.Mitral atresia :

A severe form of complex CHD because the atretic part is the main chamber (left ventricle). It has a spectrum of severity; the most severe is HLHS where hypoplasia involves the mitral valve, LV, AV and aortic arch. A less severe form occurs if the AV and aortic arch are preserved
Presentation: the severe form as in (Scenario 4 A) presents similar to severe coarctation. Less severe forms present with CHF and mild cyanosis.
Management: Staged palliation depends on the size of the aortic valve and arch.

Double outlet RV

As the name implies, both aorta and PA arise from the right ventricle. DORV is an echocardiographic diagnosis and clinically it can mimic many other CHD. It is classified into 4 types according to the position of the VSD in relation to the great arteries

A. **Simple DORV:** Subaortic VSD: Appears on echo as a large malaligned VSD with overriding of the aorta by >%50%. Clinically, this type behaves like a simple VSD causing CHF and *no cyanosis*. Management: VSD closure which should be considered early as the large malaligned VSD does not usually close spontaneously.
B. **DORV with sub pulmonary VSD: The great arteries are transposed** so the patient behaves like TGA with CHF and *severe cyanosis*. A common association is aortic arch hypoplasia and coarctation which is known as Taussig Bing deformity. Management: each patient is discussed and management plan individualized according to the anatomy. One stage repair (arterial switch with VSD closure and arch repair) is an extensive operation but can be done in some centers. Staged repair by PA banding with or without BT shunt and arch repair which also carries some risks.
C. **DORV with doubly committed VSD:** CHF with mild cyanosis: Management depends on echo findings can be simple VSD closure or more complex tunnel repair.
D. **DORV with non-committed VSD:** CHF with mild cyanosis. Management depends on echo findings.

Other conditions:

TGA/VSD/PS: Behave like TOF
AVSD/TOF: Behaves like TOF

Congenitally Corrected (L)TGA:

The ventricles are inverted (LV on the right and RV on the left) ,the aorta is connected to the RV and PA to LV therefore there is atrioventricular and ventriculo-arterial discordance i.e double discordance.The relation of the great arteries to each other is abnormal ; Ao is anterior and to the *left* of PA (instead of PA being anterior and to the left of Ao) therefore the name of ‘L’ TGA in contrast to ‘D’ TGA where the aorta is anterior and to the right of PA.

Patho-physiologically: pink blood from pulmonary veins enter the left atrium then the RV to aorta and blue blood to right atium then LV to pulmonary artery so the patient can be clinically completely normal.

However: there are many problems associated with Congenitally Corrected TGA

1. The RV often fails as it is pumping against the systemic aortic pressure.
2. The TV often fails (for the same reason) causing severe TR which presents with *pulmonary edema* (left AVV).
3. Many patients have other associated lesions like VSD and PS.
4. Rhythm disturbances such as heart block and SVT are common. Heart block can be spontaneous or precipitated by interventions like cath and surgery.

Management:

Depends on the associated anatomy and complications. Surgical procedures like VSD closure carry higher risk for L TGA patients than the other patients. Anatomical repair is often done by doing a ‘double switch’ operation i.e. both atrial and arterial switch. Physiological /conventional repair means that surgery is done only to repair the associated defects leaving the RV as the systemic ventricle. Both operations have their pros and cons and the treatment of each patient should be tailored to the anatomy and pathophysiology considering the risk of heart block

Congenital Heart Surgery

Congenital heart surgery had improved dramatically in the last 40 years due to the following:

1. Improved early accurate diagnosis due to improved echo technology and fetal echo.
2. Improved early medical treatment with introduction of prostaglandin and intensive care management.
3. Improved intraoperative care: cardiopulmonary bypass and surgical techniques.
4. Improved postoperative care: specialized personnel and equipment

Surgical repair:

Many defects can be repaired in one stage like VSD, ASD, AVSD, TOF, APVD, CoAo and PS. The timing of complete repair depends on the type of lesion, the weight of the patient and the expertise and facilities of the surgeon and centre.

Surgical Palliation:

Palliation can be done in 2 situations:

1. As a step before complete repair like Blalock-Taussig shunt for TOF and pulmonary artery band for large VSD when the patient is symptomatic but the weight or centre facilities do not allow complete repair or the size of the pulmonary artery branches (in TOF) or size and number of VSDs (in large/multiple VSDs) prohibit one stage repair .
2. As a final surgery: when there is functionally single ventricle or a very complex un-repairable anatomy *staged palliation* is performed as a final surgery.

The most common palliative procedures are the Blalock-Taussig shunt (BTS) and PA banding. BTS is done for TOF and TOF-like conditions while PA band is done for large VSD when complete repair is not feasible. The 2 procedures are non- open cardiac surgery (no cardiopulmonary bypass) usually done from the lateral thoracotomy approach.

Staged Palliation:

These procedures are known as the ‘single ventricle pathway’, the Glenn/Fontan pathway or the cavo-pulmonary connections. They were first described by Fontan in 1971 for patients with tricuspid atresia and since that time a series of modifications have been described. The whole mark of this procedure is to direct the systemic venous return directly to the lungs (bypassing the heart) and to leave the ventricle as the pumping chamber to the body. There are two main operations: the Glenn anastomosis (superior vena cava to pulmonary artery) which is done at the age of 4-6 months and the Fontan anastomosis (inferior vena cava to pulmonary artery) which is done later at around 3 years of age. (Figure85).

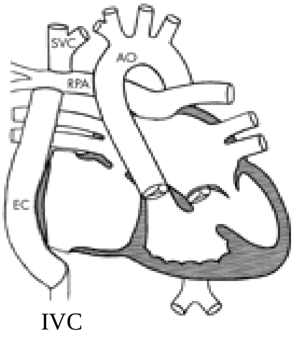


Figure 85: A diagram showing a heart with tricuspid atresia and Glenn and Fontan anastomoses.
SVC: superior vena cava
RPA: right PA
EC: External conduit

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Pre-Glenn Palliation:

Patients with functionally single ventricle can become symptomatic in the neonatal period with cyanosis(when CHD is associated severe PS) or with CHF and mild cyanosis (when there is no PS). At this stage the Glenn operation cannot be done as the pulmonary pressure and vascular resistance is still high therefore the blood flow from the SVC to PA will be impaired. Consequently these patients should have either a Blalock-Taussig shunt (for severe cyanosis) or a PA band (for CHF) (Figure 86)

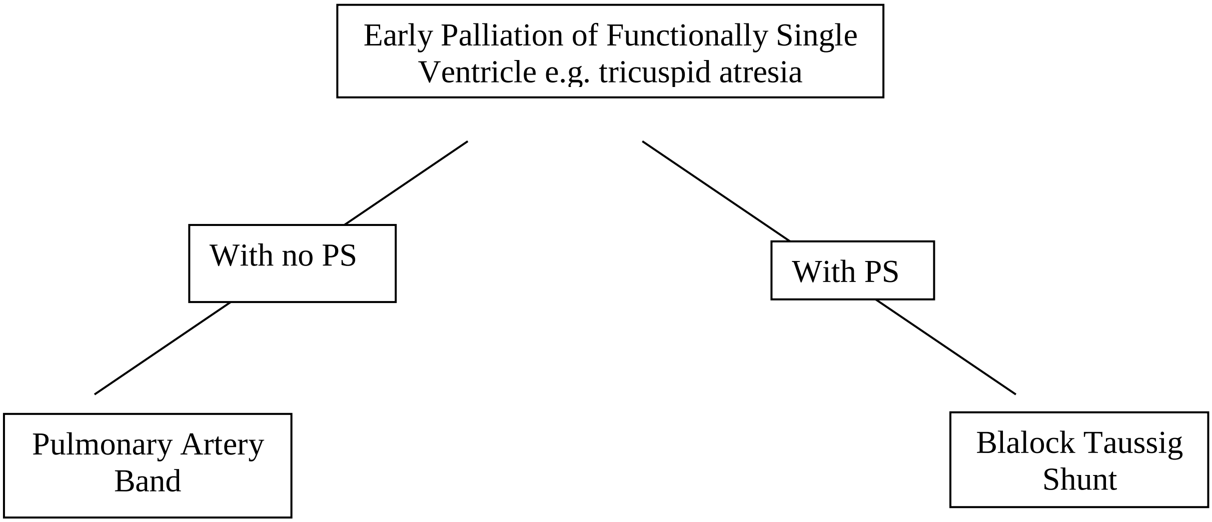

Early Palliation of Functionally Single Ventricle e.g. tricuspid atresia
With no PS
With PS
Pulmonary Artery Band
Blalock Taussig Shunt

Figure86
The types of early palliation (pre-Glenn) for patients with functionally single ventricle.

Hemodynamics of the Cavo-pulmonary Connection (CPC):
As caval veins, which have a low pressure (mean of 7-10 at most), will be connected to the PA which normally has a mean pressure of 15 mmHg, the blood flow in the CPC is limited by the pressure gradient across these two vessels. Any factor that leads to increase of the PA pressure will lead to failure of the CPC e.g increased left to right shunt, AVV regurgitation, ventricular dysfunction and small PA branch diameter.

Prerequisites to CPC:

1. Adequate PA branch size
2. Minimal/no AVV regurgitation
3. Good ventricular function.
4. Mean PA pressure <15 mmHg.

Long term outcome of CPC:

The survival rates are improving with more understanding of the CPC pathophysiology and refinements of the surgical techniques. A 10 year survival of around 80% has been reported, in some studies survival rates were up to 90% at 10years. The 20 year survival is reported to be 70%. There are a number of associated morbidities in the form of arrhythmias, thromboembolism and CHF. Many patients need re interventions (surgical or trans cathter) for AVV regurgitation or closure of fenestration and collateral vessels. There is a definite need for regular meticulous follow up by the paediatric cardiologist or adult CHD specialist.

Heterotaxy Syndromes (Isomerism)

Isomerism means abnormal 'sidedness'.

Normally the right side of the body is different from the left, e.g the left lung has 2 and right lung 3 lobes, the liver is on the right while the heart and spleen are on the left, etc. In left isomerism there is abnormal left sidedness where the both sides look like the left side (polysplenia) and in right isomerism both sides look like the right side (asplenia). Isomerism is often associated with a central liver, mesocardia or dextrocardia and complex congenital heart disease. (Figure87)

Isomerism : abnormal sidedness.
Left isomerism : Bilateral left sidedness (polysplenia)
Right isomerism : Bilateral right sidedness (asplenia)

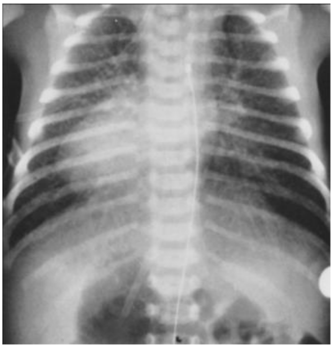


Figure 85:
CXR of a patient with isomerism showing dextrocardia with central liver.

Associations:

- Both left and right isomerism are associated with complex CHD, commonly dextrocardia, AVSD, APVD, interrupted IVC (more in left isomerism).
- Conduction abnormalities due to abnormal position of the AV node.
- Splenic dysfunction.
- Usually unfavorable surgical course.

Echocardiographic features:

- The abdominal situs (i.e the position of the main organs) is determined from the subcostal situs view. In the normal situs (situs solitus) the aorta lies to the left and IVC to the right of the spine. In isomerism the situs is referred to as : situs ambiguous which has 2 patterns:
- In left isomerism the IVC is interrupted and the lower body venous blood is carried by the azygous vein which passes parallel to aorta to connect to the superior vena cava. In the situs view the aorta and azygous vein are seen in one view on the same side of the spine. Hepatic veins are seen draining to the right atrium directly (no IVC) and the liver is often central.

- In right isomerism: aorta and IVC are to the same side of the spine. The liver is often central (Figure 86).

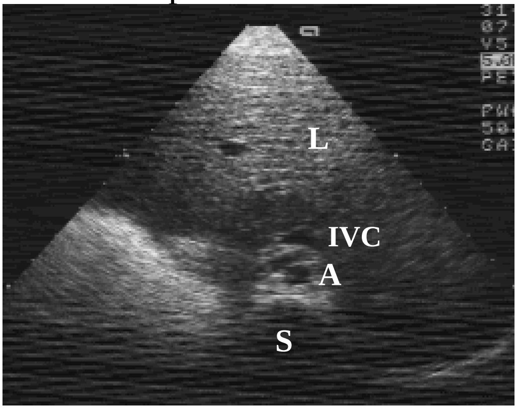


Figure 86
Echocardiographic short axis (situs view) showing the aorta (A) and inferior vena cava (IVC) to the same side of the spine (S), the liver (L) lies to the right of the spine. This patient has right atrial isomerism.

Management Issues:

- Complete repair or palliation can be done in patients with isomerism depending on their anatomical and patho physiological conditions. Traditionally the outcome of repair or palliation is less successful in patients with hetertotaxy when compared with other patients. There had been some reports of improving outcome but patients with AVV regurgitation, APVD and non-sinus rhythm still constitute a high risk category.
- Ultrasound to locate the spleen needs to be done as asplenia and polysplenia can be present. If the spleen is not found this is called anatomical asplenia.
- Functional asplenia: the spleen can be present but not functioning. This is investigated by peripheral blood film for Howell-Jolly bodies, when present in the blood film they indicate functional asplenia. Splenic isotope scan can also be done to look for splenic function. If there is functional or anatomical asplenia the patient should be started on prophylactic oral penicillin.

Further Readings:

1.Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax

1971;26(3):240-248.

2.Lars Idorn. Univentricular Heart – Management and Prognosis.Cong Cardio Today 2010 ;(8):1-6.

3.Driscoll DJ. Long-term results of the Fontan operation. Pediatr Cardiol 2007;28(6):438-442.

4.Shin'oka T, Kurosawa H, Imai Y, Aoki M, Ishiyama M, Sakamoto T, Miyamoto S, Hobo K, Ichihara Y. Outcomes of definitive surgical repair for congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections: risk analyses in 189 patients. J Thorac Cardiovasc Surg. 2007 May;133(5):1318-28, 1328.e1-4. Epub 2007 Mar 21.

5. Anagnostopoulos PV, Pearl JM, Octave C, Cohen M, Gruessner A, Wintering E, Teodori MF. Improved current era outcomes in patients with heterotaxy syndromes. Eur J Cardiothorac Surg. 2009 May;35(5):871-7

Ethical Issues in Paediatric Cardiology

Physicians play a role in their patients’ lives that is literally not ‘the healing role’; in fact healing comes from ‘God’ and not from humans. Therefore the role of physicians is to help their patients: help them find out problems and try to solve them. A good communication is highly needed so the ***Golden Rules*** are:

1. Give your patients your full attention and keep parents comfortable while you examine their child.
2. Smile: a smile from the doctor provides more help than medicine.
3. Give time and eye to eye contact to your patient and his family.
4. Always sit and explain to parents (and child) the problems in simple words and answer their queries.
5. Remind them about Quraan and Duaa for healing.

Breaking news to the family:

As in all other branches of medicine breaking news needs more art than science particularly when this is about a serious CHD. This can be even more difficult as it is often done in the neonatal period when parents had not yet come back to their steady state. On the other hand, it can be done when the child is not experiencing any symptom (as when the patient is referred due to a murmur) so parents' apprehension would be greatly affected.

How can we make this event smooth?

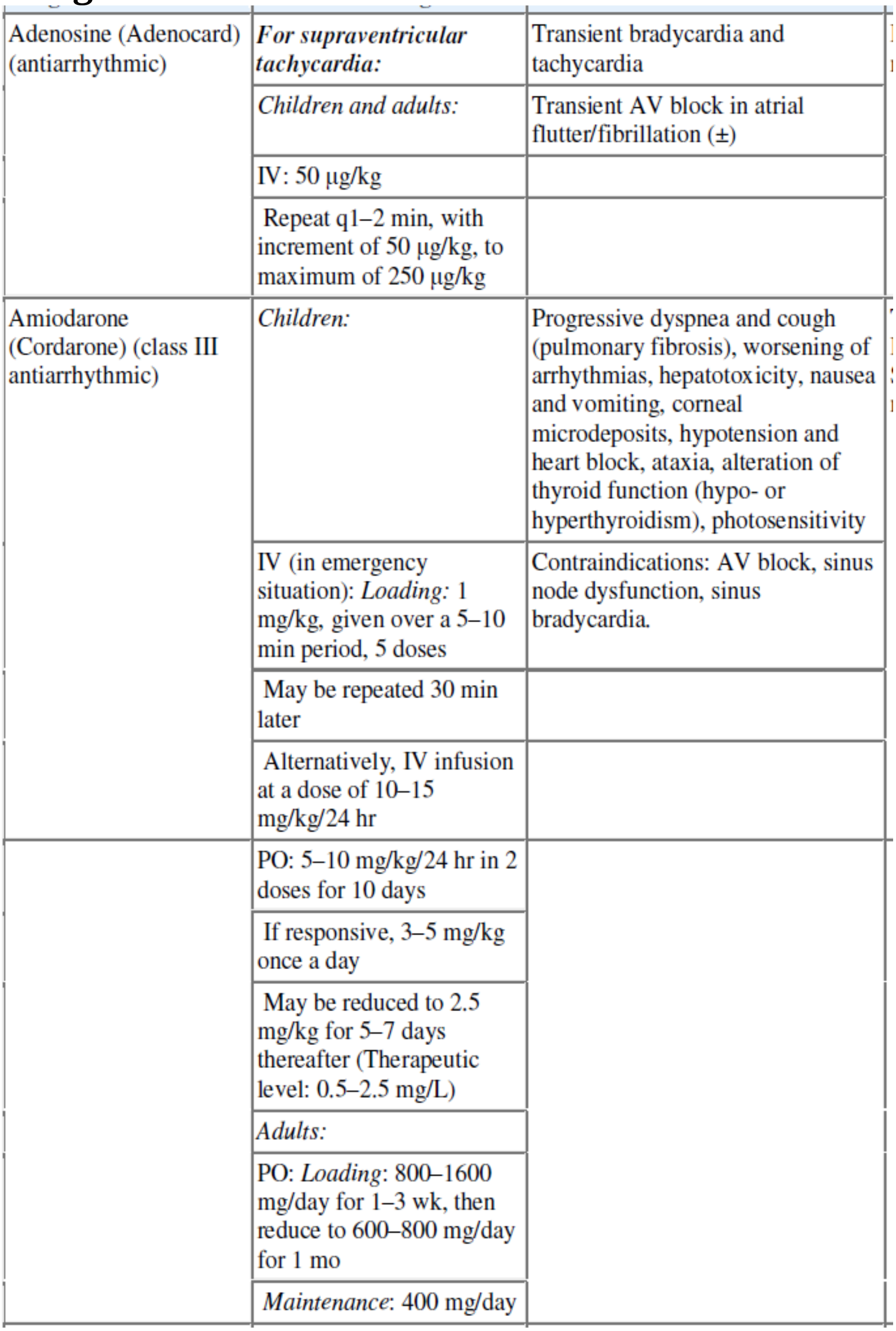
- After complete evaluation of the patient, sit with the parents in a quiet place.
- Counseling is ideally done with the parents; far relatives are usually not involved *unless* the parents wish to do so. Closer relatives and friends can be present (upon parents' wish) to support parents.
- Try to find out if the father wishes that the mother be involved in the initial counseling. It is better to speak to both parents.
- Try to use sirmple non-medical terms to explain the disease using diagrams.
- Explain the options available: when surgery is available in another centre tell the parents about this option.
- Show that you will continue to care for the patient medically when surgery is not an option.
- Give enough time, eye to eye contact and a lot of sympathy.
- Offer a second session for counseling if parents are not able to grasp information from the first time, this is particularly important for complex CHD..
- Try to answer all questions raised by parents.

Inoperable/ End-Stage Disease:

Many complex CHD are inoperable or need very advanced facilities. . Similarly cardiomyopathies often lead to end-stage CHF which does not respond to medical treatment. Families with poor socio-economic status sometimes easily accept this fact; however we, as physicians should do our best before we get to this conclusion:

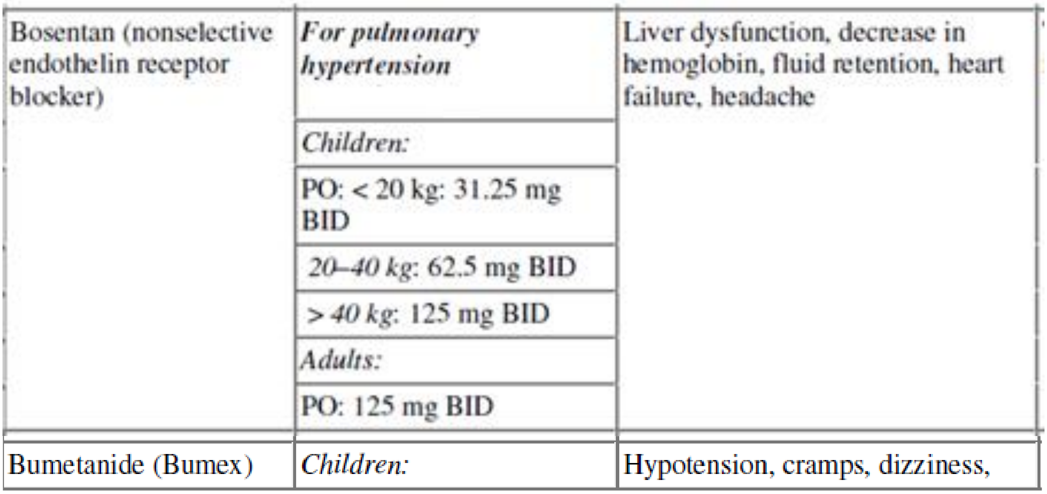
1. Make sure that your anatomical diagnosis is correct, if in doubt bring the patient for another session or consult an experienced colleague.
2. Make sure that you know the most recent treatment available if you are not sure, search the literature.
3. A second opinion is warranted: this can easily be obtained by e mail from colleagues working in more advanced centres.
4. Make sure you give all the options available locally and abroad; however keep the balance with parents' social and economical state.
5. Continue following up the patient and supporting the family.

Appendix I
Commonly Prescribed Medications:



Drug Name	Dose	Side Effects
Adenosine (Adenocard) (antiarrhythmic)	***For supraventricular tachycardia:***	Transient bradycardia and tachycardia
	Children and adults:	Transient AV block in atrial flutter/fibrillation (±)
	IV: 50 µg/kg	
	Repeat q1–2 min, with increment of 50 µg/kg, to maximum of 250 µg/kg	
Amiodarone (Cordarone) (class III antiarrhythmic)	*Children:*	Progressive dyspnea and cough (pulmonary fibrosis), worsening of arrhythmias, hepatotoxicity, nausea and vomiting, corneal microdeposits, hypotension and heart block, ataxia, alteration of thyroid function (hypo- or hyperthyroidism), photosensitivity
	IV (in emergency situation): *Loading:* 1 mg/kg, given over a 5–10 min period, 5 doses	Contraindications: AV block, sinus node dysfunction, sinus bradycardia.
	May be repeated 30 min later	
	Alternatively, IV infusion at a dose of 10–15 mg/kg/24 hr	
	PO: 5–10 mg/kg/24 hr in 2 doses for 10 days	
	If responsive, 3–5 mg/kg once a day	
	May be reduced to 2.5 mg/kg for 5–7 days thereafter (Therapeutic level: 0.5–2.5 mg/L)	
	Adults:	
	PO: *Loading*: 800–1600 mg/day for 1–3 wk, then reduce to 600–800 mg/day for 1 mo	
	Maintenance: 400 mg/day	

Acetylsalicylic acid (aspirin)	***Antiplatelet therapy:*** *Children:* PO: 3–5 mg/kg every day	Rash, nausea, hepatotoxicity, GI bleeding, bronchospasm, GI distress, tinnitus
	Antipyretic/analgesic: PO: 10–15 mg/kg/dose, q4–6hr (Max. 4 g/24 hr) ***Anti-inflammatory:*** *Children:* PO: 80–100 mg/kg/24 hr in 4 doses	Contraindications: hepatic failure, blooding disorder, hypersensitivity, children <16 yr old with chickenpox or flu symptoms (due to the association with Reye's syndrome)
Atenolol (Tenormin) (β_1-adrenoceptor blocker, antihypertensive, antiarrhythmic)	*Children:* PO: 1–2 mg/kg/24 hr *Adults:* PO:50 mg once a day for	CNS symptoms (dizziness, tiredness, depression), bradycardia, postural hypotension, nausea and vomiting, rash, blood dyscrasias (agranulocytosis, purpura)
	1–2 wk (alone or with diuretic for hypertension) May increase to 100 mg once a day	
Atorvastatin (Lipitor) (lipid-lowering agent, ("statin"), HMG-CoA reductase inhibitor)	*Children:* PO: Starting dose 10 mg QD, for 4–6 wk Increase to 20 mg and 40 mg as needed. (Adult max. dose 80 mg/day)	Headache, constipation, diarrhea, elevated liver enzymes, rhabdomyolysis, myopathy



Bosentan (nonselective endothelin receptor blocker)	***For pulmonary hypertension***	Liver dysfunction, decrease in hemoglobin, fluid retention, heart failure, headache
	Children:	
	PO: < 20 kg: 31.25 mg BID	
	20–40 kg: 62.5 mg BID	
	> 40 kg: 125 mg BID	
	Adults:	
	PO: 125 mg BID	
Bumetanide (Bumex) (loop diuretic)	*Children:*	Hypotension, cramps, dizziness, headache, electrolyte losses (hypokalemia, hypocalcemia, hyponatremia, hypochloremia), metabolic alkalosis
	PO, IM, IV: >6 mo: 0.015–0.1 mg/kg/dose, QD-QOD (max. dose: 10 mg/24 hr)	
	Adults:	
	PO: 0.5–2 mg/dose, QD-BID MI, IV: 0.5–1 mg over 1–2 min, q2–3hr prn.	

Captopril (Capoten) (angiotensin-converting enzyme inhibitor, antihypertensive, vasodilator)	*Children:*	Neutropenia/agranulocytosis, proteinuria, hypotension and tachycardia, rash, taste impairment, small increase in serum potassium levels (±)
	PO: *Newborn:* 0.1–0.4 mg/kg/24 hr, in QD-QID;	
	Infant: Initially 0.15–0.3 mg/kg/dose. Titrate upward if needed. Max. dose 6 mg/kg/24 hr, QD-QID.	
	Child: Initially 0.3–0.5 mg/kg/dose q8hr. Titrate upward if needed. Max. 6 mg/kg/24 hr, BID-QID.	Evidence of fetal risk if given during second and third trimesters (same with all other ACE inhibitors)
	Adolescent and adult:	
	Initially 12.5–25 mg/dose, BID-TID. Increase weekly if needed by 25 mg/dose to max. dose 450 mg/24 hr.	
	Smaller dose in renal impairment	
Carnitine (Carnitor)	*Children:*	Nausea/vomiting, abdominal cramp, diarrhea
	PO: 50–100 mg/kg/24 hr, q8-12hr	
	Increase slowly as needed (max. 3 g/day)	
	Adults:	
	PO: 330 mg–1 g/dose, BID-TID	

Carvedilol (Coreg, Coreg Tiltabs) (nonselective β-adrenergic blocker)	*Children:* PO: Initial 0.09 mg/kg/dose, q12hr Increase gradually to 0.36 and 0.75 mg/kg as tolerated to adult max. dose of 50 mg/24 hr. *Adults:* PO: 3.125 mg, BID for 2 wk; increase slowly to a max. dose of 25 mg BID as needed (for heart failure)	Dizziness, hypotension, headache, diarrhea, rarely AV block.

Chloral hydrate (Noctec) (sedative, hypnotic)	***As sedative:*** *Children:* PO, PR: 25 mg/kg/dose q8hr *Adults:* PO, PR: 250 mg/dose q8hr ***As hypnotic:*** *Children:* PO, PR: 50–75 mg/kg/dose *Adults:* PO, PR: 500-2000 mg/dose	Mucous membrane irritation (laryngospasm if aspirated), GI irritation, excitement/delirium (contraindicated in hepatic and renal impairment)
Chlorothiazide (Diuril) (diuretic)	*Children:* PO: 20–40 mg/kg/24 hr, BID	May increase serum calcium, ↑bilirubin, glucose, and uric acid.
	Adults:	Hypochloremic alkalosis,

Cholestyramine (Questran, Prevalite) (cholesterol-lowering agent)	*Children:* PO: 250–1500 mg/kg/24 hr in 2–4 doses *Adults:* PO: *Starting:* 1 packet (or scoopful) of Questran powder or Questran Light 1–2 times/day *Maintenance:* 2–4 packets or scoopfuls/day in 2 doses (or 1–6 doses) *Maximum:* 6 packets/day	Constipation and other GI symptoms, hyperchloremic acidosis, bleeding
Clopidogrel (Plavix) (antiplatelet agent)	*Children:* PO: 1 mg/kg/day to max. (adult dose) of 75 mg/day *Adults:*	Bleeding, especially when used with aspirin, neutropenia or agranulocytosis, abdominal pain, constipation, rash, syncope,
Diazoxide (Hyperstat, Proglycem) (peripheral vasodilator)	***For hypertensive crisis:*** *Children and adults:* IV: 1–3 mg/kg (maximum 150-mg single dose), repeat q5–15 min, titrate to desired effect	Hypotension, transient hyperglycemia, nausea and vomiting, sodium retention (CHF±)

Digoxin (Lanoxin) (cardiac glycoside)	*Children:* ***TDD:*** PO: Premature infant: 20 µg/kg; Full-term newborn: 30 µg/kg; Child 1 mo–2 yr: 40-50 µg/kg; Child >2 yr: 30–40 µg/kg IV: 75%–80% of PO dose ***Maintenance:*** PO: 25%–30% of TDD/day in 2 doses *Adults:* PO: *Loading:* 8–12 µg/kg *Maintenance:* 0.10–0.25 mg/day	AV conduction disturbances, arrhythmias, nausea and vomiting (see Box 30-1 for ECG changes)
Digoxin immune Fab (Digibind) (digoxin antidote)	*Infants and children:* IV: 1 vial (40 mg) dissolved in 4 mL H_2O, over 30 min *Adults:* IV: 4 vials (240 mg)	Allergic reaction (rare), hypokalemia, rapid AV conduction in atrial flutter
Dobutamine (Dobutrex)	*Children:*	Tachyarrhythmias, hypertension,
(β_1-adrenergic stimulator)	IV: 2–15 µg/kg/min in D5W or NS (incompatible with alkali solution) *Adults:* IV: 2.5–10 µg/kg/min (maximum 40 µg/kg/min)	nausea and vomiting, headache (contraindicated in IHSS and atrial flutter/fibrillation)

Dopamine (Intropin, Dopastat) (natural catecholamine inotropic agent)	*Children:*	Tachyarrhythmias, nausea and vomiting, hypotension or hypertension, extravasation (tissue necrosis [treat with local infiltration of phentolamine])
	IV: Effects are dose dependent:	
	2–5 µg/kg/min—increases RBF and urine output	
	5–15 µg/kg/min—increases heart rate, cardiac contractility and cardiac output.	
	>20 µg/kg/min—α-adrenergic effects with decreased RBF (±) (Incompatible with alkali solution)	
Enalapril, Enalaprilat (Vasotec) (ACE inhibitor, vasodilator)	*Children:*	Hypotension, dizziness, fatigue, headache, rash, diminishing taste, neutropenia, hyperkalemia, chronic cough
	PO: 0.1 mg/kg/dose QD or BID (maximum 0.5 mg/kg/day)	Evidence of fetal risk if given during second and third trimesters (same with all other ACE inhibitors)
	Adults:	
	For CHF:	
	PO: Start with 2.5 mg once or twice daily (usual range 5–20 mg/day)	
	For hypertension:	
	PO: Start with 5 mg once a day (usual dose 10–40 mg/day)	

Epinephrine (Adrenalin) (α-, β_1-, and β_2-adrenergic stimulator)	*Children:*	Tachyarrhythmias, hypertension, nausea and vomiting, headache, tissue necrosis (±)
	IV: 1:10,000 sol—begin with 0.1 μg/kg/min; increase to 1 μg/kg/min to achieve desired effect	

Esmolol (Brevibloc) (β_1-selective adrenergic blocking agent, antihypertensive. class II antiarrhythmic)	*Children:*	Bronchospasm, CHF, hypotension, nausea/vomiting
	IV infusion:	
	Loading dose: 100– 500 µg/kg over 1 min.	
	Maintenance: 25–100 µg/kg/min as infusion, increase by 50 µg/kg to a maximum of 300 µg/kg/min	
	(Usual maintenance dose 50–500 µg/kg/min)	
Ethacrynic acid (Edecrin) (loop diuretic)	*Children:*	Dehydration, hypokalemia, prerenal azotemia, hyperuricemia, eighth cranial nerve damage (deafness), abnormal LFT, agranulocytosis or thrombocytopenia, GI irritation, rash
	PO: 25 mg/dose once a day (maximum 2–3 mg/kg/24 hr)	
	IV: 1 mg/kg/dose	
	Adults:	
	PO: 50–100 mg once a day (maximum 400 mg)	

	IV: 0.5–1 mg/kg/dose or 50 mg/dose	
Fentanyl (Sublimaze, Duragesic, Fentanyl Oralet) (narcotic analgesic)	***For sedation:***	Respiratory depression, apnea, rigidity, bradycardia
	Children:	
	IV: 1–3 yr: 2–3 µg/kg/dose; 3–12 yr: 1–2 µg/kg/dose; >12 yr: 0.5–1 µg/kg/dose; may repeat q30–60 min	
	PO (Fentanyl Oralet) for sedation:	
	10–15 µg/kg/dose. (Max. 400 µg/dose)	
	Adults:	
	IV: 50–100 µg/dose	
Flecainide (Tambocor) (Class IC antiarrhythmic)	***For sustained ventricular tachycardia:***	Worsening of HF, bradycardia, AV block, dizziness, blurred vision, dyspnea, nausea, headache, increased PR and QRS duration
	Children:	Reserve for life-threatening cases.
	PO: Initial 1–3 mg/kg/day, q8hr.	

	Usual range: 3–6 mg/kg/day, q8hr	
	Monitor serum level to adjust dose if needed.	
	[Therapeutic trough level after 2–3 days of continuous dosing: 0.2–1 mg/L]	
	Adults:	
	PO: 100 mg, q12hr.	
	May increase by 50 mg q12hr every 4 days to max. dose of 600 mg/24 hr	
Fludrocortisone acetate (Florinef, Fluohydrisone) (corticosteroids)	***For syncopal episodes:***	Hypertension, hypokalemia, acne, rash, bruising, headache, GI ulcers, and growth suppression.
	Children:	Weight gain (1–2 kg in 2–3 wk)
	PO: 0.1 mg/dose, once or 2 times a day	

	Adults:		
	PO: 0.2 mg/dose, once or 2 times		
Furosemide (Lasix, Furomide) (loop diuretic)	*Children:*	Hypokalemia, hyperuricemia, prerenal azotemia, ototoxicity, rarely bloody dyscrasias, rash	Oral liquid: 10 mg/mL 40 mg/5 mL Tab: 20, 40, 80 mg Inj: 10 mg/mL
	IV: 0.5–2 mg/kg/dose 2–4 times/day		
	PO: 1–2 mg/kg/dose 1–3 times/day prn (maximum 6 mg/kg/dose)		
	Adults:		
	IV: 20–40 mg/dose 2–4 times/day		
	PO: 20–80 mg/dose 1–4 times/day prn		
Heparin (anticoagulant)	*Infants and children:*	Bleeding	Inj: 1000, 2500, 5000, 7500, 10,000 U/mL
	IV: Initial: 50 U/kg IV bolus	Antidote: protamine sulfate (1 mg per 100 U heparin in previous 4 hr)	
	Maintenance: 10–25 U/kg/hr as IV infusion, or 50–100 U/kg q4hr IV		
	[Adjust dose to give PTT 1.5–2.5 times control, 6–8 hr after IV infusion (or 3.5–4 hr after injection)]		

	Adults:	
	IV: *Initial:* 10,000 U IV inj	
	Maintenance: 5000–10,000 U q4-6hr	
	IV drip: *Initial dose:* 5000 U followed by 20,000–40,000 U/24 hr	

Hydralazine (Apresoline) (peripheral vasodilator, antihypertensive)	*Children:*	Hypotension, tachycardia and palpitation, lupus-like syndrome with prolonged use (fever, arthralgia, splenomegaly and positive LE-cell preparation), blood dyscrasias
	IM, IV: 0.15–0.2 mg/kg/dose (for emergency); may be repeated q4–6hr	
	PO: 0.75–3 mg/kg/day in 2–4 doses	
	Adults:	
	IM, IV: 20–40 mg/dose	

	(for emergency); repeat prn	
	PO: Start with 10 mg 4 times/day for 3–4 days, increase to 25 mg 4 times/day for 3–4 days, then up to 50 mg 4 times/day	
Hydrochlorothiazide (HydroDIURIL, Esidrix, Hydro-Par, Oretic) (thiazide diuretic)	*Children:*	Same as for chlorothiazide
	PO: 2–4 mg/kg/24 hr in 2 doses	
	Adults:	
	PO: 25–100 mg/24 hr, single or divided doses	
	May be given intermittently	

Indomethacin (Indocin) (nonsteroidal anti-inflammatory, antipyretic agent, PG synthesis inhibitor)	***For PDA closure in premature infants:***	GI or other bleeding, GI disturbances, renal impairment, electrolyte disturbances (decreased sodium and increased potassium levels)
	IV: *<48 hr:* 0.2, 01, and 0.1 mg/kg/dose, q12–24 hr.	
	2–7 days: 0.2, 0.2, and 0.2 mg/kg/dose, q12–24 hr	
	>7 days: 0.2, 0.25, and 0.25 mg/kg/dose, q12–24 hr	
	to desired effect (incompatible with alkali solution)	
Ketamine (Ketalar) (dissociate anesthetic)	*Children:*	Hypertension/tachycardia, respiratory depression or apnea, CNS symptoms (dream-like state, confusion, agitation)
	IM: 8–12 mg/kg	
	Repeat smaller doses q30min prn	
	IV: 2–3 mg/kg/dose	
	Repeat smaller doses q30min prn	
Labetalol (Normodyne, Trandate) (α- and β-adrenergic antagonist)	*Children:*	Orthostatic hypotension, edema, CHF, bradycardia.
	PO: Initial 4 mg/kg/24 hr, BID	Contraindicated in asthma.
	Max. 40 mg/kg/24 hr.	
	IV: (for hypertensive emergency)	
	Initial 0.1–1 mg/kg/dose q10min, prn (Max. 20 mg/dose)	

Lidocaine (Xylocaine) (class IB antiarrhythmic)	*Children:*	Seizure, respiratory depression, CNS symptoms (anxiety, euphoria or drowsiness), arrhythmias, hypotension or shock
	IV: Loading: 1 mg/kg/dose q5–10min prn	
	Maintenance: 30 μg/kg/min IV drip (range 20–50 μg/kg/min)	
	Adults:	
	IV: *Loading:* 1 mg/kg/dose q5min	
	Maintenance: 1–4 mg/min IV drip	

Lisinopril (Zestril, Prinivil) (ACE inhibitor, antihypertensive)	***For hypertension***	Dry nonproductive cough, rash, hypotension, hyperkalemia, angioedema, rarely marrow depression
	Children:	
	PO: Initial 0.07 mg/kg/day, May increase up to 5 mg/day (max, 0.6 mg/kg/day or 40 mg/day)	
	Adults:	Evidence of fetal risk if given during second and third trimesters (same with all other ACE inhibitors)
	PO: Initial 10 mg QD	
	May increase upward as needed to max. 80 mg/day	
Losartan (Cozaar) (angiotensin receptor blocker)	***For hypertension***	Hypotension, dizziness, nasal congestion, muscle cramps.
	Children:	
	PO: 0.7 mg/kg/24 hr, QD-BID, up to 50 mg/24 hr	
	Adults:	
	PO: Initial dose 50 mg, QD with maximum dose of 100 mg QD	

Metoprolol (Lopressor) (β_1-adrenoceptor blocker)	*Children >2 yr:*	CNS symptoms (dizziness, tiredness, depression), bronchospasm, bradycardia, diarrhea, nausea and vomiting, abdominal pain
	PO: Initial 0.1–0.2 mg/kg/dose BID	
	Gradually increase to 1–3 mg/kg/24 hr.	
	Adults:	
	PO: 100 mg/day in 1–3 doses initially	
	May increase to 450 mg/24 hr in 2–3 doses	
	(Usual dose 100–450 mg/24 hr)	
	(Usually used with hydrochlorothiazide 25–100 mg/day)	
Metolazone (Zaroxolyn, Diulo, Mykrox) (thiazide-like diuretic)	*Children:*	Electrolyte imbalance, GI disturbance, hyperglycemia, bone marrow depression, chills, hyperuricemia, hepatitis, rash.
	PO: 0.2–0.4 mg/kg/24 hr QD-BID	
	Adults:	
	PO: For hypertension: 2.5–5 mg QD	May be more effective than thiazide diuretics in impaired renal function
	For edema: 5–20 mg, QD	

Morphine sulfate (narcotic analgesic)	*Children:*	CNS depression, respiratory depression, nausea and vomiting, hypotension, bradycardia
	SC, IM, IV: 0.1-0.2 mg/kg/dose q2-4hr (maximum 15 mg/dose)	
	Adults:	
	SC, IM, IV: 2.5-20 mg/dose q2-6hr pm	
Naloxone (Narcan) (narcotic antagonist)	*Children:*	Ventricular arrhythmia, pulmonary edema (±), nausea and vomiting, seizure
	IM, IV: 5-10 µg/kg/dose q2-3min for 1-3 doses prn (may need 5-10 doses)	
	Adults:	
	IM, IV: 0.4-2 mg/dose q2-3min for 1-3 doses prn	
Nifedipine (Procardia, Adalat) (calcium channel blocker)	***For hypertrophic cardiomyopathy:***	Hypotension, peripheral edema, CNS symptoms (headache, dizziness, weakness), nausea
	Children:	
	PO: 0.6-0.9 mg/kg/24 hr in 3-4 doses	
	For hypertension:	
	Children:	

	PO: 0.25-0.5 mg/kg/24 hr in 1-2 doses (max. 3 mg/kg/24 hr up to 120 mg/24 hr)		
	Adults:		
	PO: Initially 10 mg 3 times/day		
	Titrate up to 20 or 30 mg 3-4 times/day over 7-14 days		
	(Usual dose 10-20 mg 3 times/day; maximum 180 mg/day)		
Nitroglycerine (Nitrobid, Tridil, Nitrostat) (peripheral vasodilator)	*Children:*	Hypotension, tachycardia, headache, nausea and vomiting	I
	IV: 0.5-1 µg/kg/min		
	Increase 1 µg/kg/min q20min to titrate to effect (maximum 6 µg/kg/min)		

	(Dilute in D5W or NS with final concentration <400 (µg/mL; light sensitive)	
	Adults:	
	IV: Initial dose: 5 µg/min through infusion pump	
	Increase 5 µg/min q3-5min until desired effect achieved	
Nitroprusside (Nipride) (peripheral vasodilator)	*Children:*	Hypotension, sweating and palpitation, nausea and vomiting, cyanide toxicity (metabolic acidosis earliest and most reliable evidence; monitor thiocyanate level when used >48 hr and in renal failure)
	IV: 0.5-8 µg/kg/min, with BP monitoring	
	(Usual dose 2-3 µg/kg/min)	
	(Dilute stock solution	
	[50 mg] in 250-2000 mL D5W; light sensitive)	
Norepinephrine (Levophed, levarterenol) (α- and β-adrenoceptor stimulant)	*Children:*	Hypertension, bradycardia (reflex), arrhythmias, tissue necrosis (treat with phentolamine infiltration)
	IV: 0.1 µg/kg/min initially; increase dose to attain desired effect	
	Adults:	
	IV: Add 4 mL levarterenol to 1000 mL D5W, start at 2-3 mL/min (8-12 µg/min) and adjust rate	

Phenylephrine (Neo-Synephrine) (α-adrenoceptor stimulant)	***For hypotension:***	Arrhythmias, hypertension, angina
	Children:	
	IM, SC: 0.1 mg/kg/dose q1-2hr prn	
	IV: 5-10 μg/kg/dose IV bolus q10-15min or 0.1-0.5 μg/kg/min IV infusion	
	Adults:	
	IM, SC: 2-5 mg/dose ql-2hr pm	
	IV: 0.1-0.5 mg/dose IV bolus q10-15min prn	
	Start IV infusion at 100-180 μg/min; maintain at 40-60 μg/min	

Phenytoin (Dilantin) class IB antiarrhythmic)	*Children:* IV: 2-4 mg/kg/dose over 5-10 min followed by: PO: 2-5 mg/kg/day in 2-3 doses (Therapeutic level: 5-18 µg/mL for arrhythmias, 10-20 µg/mL for seizures) *Adults:* IV: 100 mg q5min (total 500 mg) PO: 250 mg 4 times for 1 day, 250 mg twice for 2 days, and 300-400 mg/day in 1-4 doses	Rash, Stevens-Johnson syndrome, CNS symptoms (ataxia, dysarthria), lupus-like syndrome, blood dyscrasias, peripheral neuropathy, gingival hypertrophy
Potassium chloride	***Supplement in diuretic therapy:*** *Children:* PO: 1-2 mEq/kg/day in 3-4 doses (0.8-1.5 mL 10% potassium chloride/kg/day, or 0.4-0.7 mL 20% potassium chloride/kg/day in 3-4 doses)	GI disturbances, ulcerations, hyperkalemia
Potassium gluconate	***Supplement in diuretic therapy:*** *Children:* PO: 1-2 mEq/kg/day in 3-4 doses or 0.8-1.5 mL/kg/day in 3-4 doses	Same as for potassium chloride

Prazosin (Minipress) (postsynaptic α-adrenergic blocker; antihypertensive)	*Children:*	CNS symptoms (dizziness, headache, drowsiness), palpitation, nausea
	PO: 5 µg/kg as a test dose, then 25-150 µg/kg/day in 4 doses	
	Adults:	
	PO: 1 mg 2-3 times/day initially	
	Increase to 20 mg/day in	
	2-4 doses	
	(Usual dose 6-15 mg/day)	

Procainamide (Pronestyl) (class IA antiarrhythmic)	*Children:*	Nausea and vomiting, blood dyscrasias, rash, lupus-like syndrome, hypotension, confusion or disorientation
	IV: *Loading:* 3-6 mg/kg/dose over 5 min repeated q10-30min (maximum 100 mg)	
	Maintenance: 20-80 (μg/kg/min by IV infusion (maximum 2 g/24 hr)	
	PO: 15-50 mg/kg/day q3-6hr (maximum 4 g/24 hr)	
	Adults:	
	IV: *Loading:* 50-100 mg/dose q5min prn	
	Maintenance: 1-6 mg/min by IV infusion	
	PO: 250-500 mg/dose q3-6hr (usual dose 2-4 g/day). Therapeutic level: 4-10 ng/mL	
Promethazine (Phenergan) (sedative, antiemetic)	***For nausea and vomiting:***	CNS stimulation, anticholinergic effects
	Children:	
	IM, PR: 0.25-0.5 mg/kg q4-6hr prn	
	Adults:	
	IM, PR: 12.5-25 mg q6hr prn	
	For sedation before surgery:	
	Children:	
	IM, PO, PR: 0.5-1 mg/kg/dose q6hr prn	
	Adults:	
	IM, PO, PR: 25-50 mg q4-6hr prn	
Propranolol (Inderal) (β-adrenoceptor blocker, class II antiarrhythmic)	***For hypertension:***	Hypotension, syncope, bronchospasm, nausea and vomiting, hypoglycemia, lethargy or depression, heart block
	Children:	
	PO: 2-4 mg/kg/24 hr in 2-4 doses (maximum 16	

	mg/kg/24 hr) *For arrhythmias:* *Children:* IV: 0.01-0.15 mg/kg/dose over 10 min (maximum 1 mg/dose) PO: 2-4 mg/kg/24 hr in 3-4 doses (maximum 16 mg/kg/24 hr) *Adults:* IV: 1 mg/dose q5min (maximum 5 mg) PO: 40-320 mg/24 hr in 3-4 doses	
Prostaglandin E_1 or alprostadil (Prostin VR) (vasodilator)	***For patency of ductus arteriosus:*** IV: Begin infusion at 0.05-0.1μg/kg/min When desired effect achieved, reduce to 0.05, 0.025, and 0.01 μg/kg/min If unresponsive, dose may be increased to 0.4 μg/kg/min	Apnea, flushing, bradycardia, hypotension, fever
Protamine sulfate (heparin antidote)	***Antidote to heparin overdose:*** IV: Each 1-mg protamine neutralizes approx 100 U heparin given in preceding 3-4 hr Slow IV infusion at rate not exceeding 20 mg/min or 50 mg/10 min Check APTT	Hypotension, bradycardia, dyspnea, flushing, coagulation problem

Sildenafil	For pulmonary Hypertension: 0.3mg/kg/dose, max 3mg/kg/dose oral, 3-6 hourly	

Simvastatin (Zocor) (lipid-lowering agent, HMG-CoA reductase inhibitor)	*Children:*	Headache, constipation, diarrhea, elevated liver enzymes, rhabdomyolysis, myopathy
	PO: Starting dose 10 mg, QD	
	Increment of 10 mg q6-8wk to max. dose of 40 mg/day as needed.	
	(Adult max. dose 80 mg/day)	
Sodium polystyrene sulfonate (Kayexalate, Kionex) (potassium-lowering agent)	***For hyperkalemia (slowly effective, taking hours to days):***	(Cation exchange resin with practical exchange rates of 1 mEq potassium per 1 g resin)
	Children:	(Note: Delivers 1 mEq sodium for each mEq of potassium removed)
	PO, NG: 1 g/kg/dose q6hr	Nausea and vomiting, constipation, severe hypokalemia (muscle weakness, confusion [monitor serum potassium levels, ECG]), hypocalcemia or hypematremia (edema)
	PR: 1 g/kg/dose q2-6hr	

	Adults:	
	PO, NG, PR: 15 g (4 level tsp) 1-4 times/day	
Spironolactone (Aldactone) (aldosterone antagonist)	*Children:*	Hyperkalemia (when given with potassium supplements), GI distress, rash, gynecomastia, agranulocytosis
	PO: 3 mg/kg/day in 1-2 doses	
	Adults:	Contraindicated in renal failure.
	PO: 50-100 mg/day in 3-4 doses (maximum 200 mg/day)	
Streptokinase (Streptase, Kabikinase) (thrombolytic agent)	***For thrombolysis:***	Potential for allergic reaction with repeated use; premedicate with acetaminophen and antihistamine, and repeat q4-6hr
	Children:	
	IV: 3500-4000 U/kg over 30 min, followed by 1000-1500 U/kg/hr, or 2000 U/kg load over 30 min followed by 2000 U/kg/hr,	
	[Duration of infusion based on response but generally does not exceed 3 days]	
	Obtain tests at baseline and q4hr: APTT, TT, fibrinogen, PT, hematocrit, platelet count	
	APTT and TT should be <2 times control	

Triamterene (Dyrenium) (potassium-sparing diuretic)	*Children:* PO: 2-4 mg/kg/24 hr in 1-2 doses. May increase up to max. 6 mg/kg/24 hr or 300 mg/24 hr *Adults:* PO: 50-100 mg/24 hr in 1-2 doses (maximum 300 mg/24 hr)	Nausea and vomiting, leg cramps, dizziness, hyperuricemia, rash, prerenal azotemia
Urokinase (Abbokinase) (thrombolytic agent)	*Children:* ***For clot lysis:*** IV: *Loading:* 4400 U/kg over 10 min	Bleeding, allergic reactions, rash, fever and chills, bronchospasm

	May repeat same dose in 15 min. Max. dose 5 mg first dose; 10 mg second dose	
	Adults:	
	IV: 5-10 mg, 10 mg second dose	
	For hypertension:	
	Children:	
	PO: 4-8 mg/kg/24 hr in 3 doses	
	Adults:	
	PO: 240-480 mg/24 hr in 3-4 doses	

Vitamin K_1 Warfarin (Coumadin, Sofarin) (anticoagulant)	***Antidote to dicumarol or warfarin:***	
	PO/IM/SC/IV: 2.5-10 mg/dose in 1 dose for correction of excessive PT from dicumarol or warfarin overdose	
	Children:	Bleeding (antidote: vitamin K or fresh-frozen plasma)
	PO: *Initial:* 0.1-0.2 mg/kg/dose QD for 2 days in evening (Max. dose 10 mg/dose)	*Increased PT response:* salicylates, acetaminophen, alcohol, lipid-lowering agents, phenytoin, ibuprofen, some antibiotics
	[Liver dysfunction, 0.1 mg/kg/day, max. 5 mg/dose]	*Decreased PT response:* antihistamines, barbiturates, oral contraceptives, vitamin C, diet high in vitamin K
	Maintenance: 0.1 mg/kg/24 hr QD.	Onset of action: 36-72 hr. and full effects in 45 days.
	Monitor INR after 5-7 days of new dosage. Keep INR at 2.5-3.5 for mechanical prosthetic valve; 2-3 for prophylaxis of DVT, pulmonary emboli.	Mode of action: inhibits hepatic synthesis of vitamin-K dependent factors (I, VII, IX, X).
	Heparin preferred initially for rapid anticoagulation; warfarin may be started	

	concomitantly with heparin or may be delayed 3-6 days	
	Adults:	
	PO: *Initial:* 5-15 mg/day for 2-5 days	
	Maintenance: 2-10 mg/day.	
	Adjust dosage based on INR	

Appendix II- A
Paediatric Basic Life Support

Unresponsive?

Shout for help

Open airway

NOT BREATHING NORMALLY?

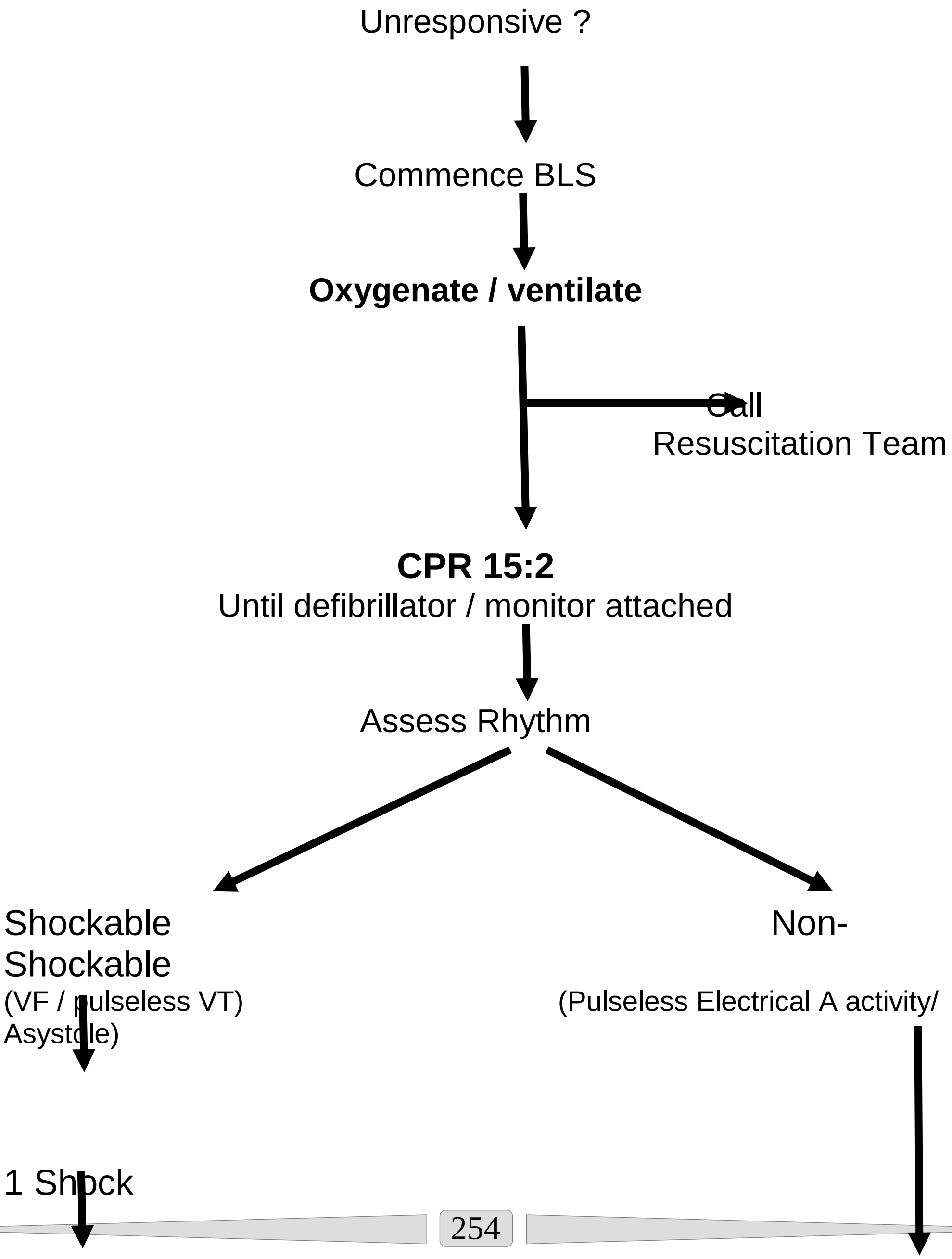
5 rescue breaths

Still unresponsive?

15 chest compressions
2 rescue breaths

After 1 minute call resuscitation team then continue CPR

Appendix II-B
Paediatric Advanced Life Support



4 J/kg o

Immediately resume immediately resume

CPR 15:2 for 2 minutes CPR 15:2 for 2 minutes

During CPR:

- Correct reversible causes*
- Check electrode position and contact
- Attempt / verify:
  - IV / IO access
  - airway and oxygen
- Give uninterrupted compressions when trachea intubated
- Give adrenaline every 3-5 minutes
- Consider: amiodarone, atropine, magnesium

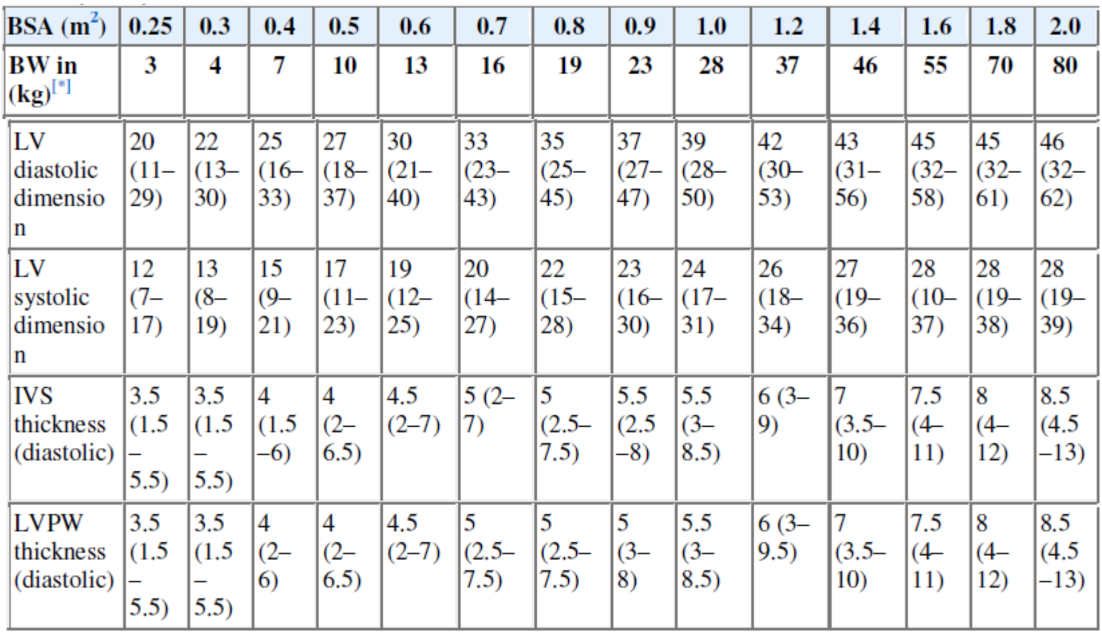
Reversible Causes

4 Fs	**4 Ts**
Hypoxia	Tension pneumothorax
Hypovolaemia	Tamponade, cardiac
Hypo/hyperkalaemia/metabolic	Toxins
Hypothermia	Thromboembolism

References:

Basic and advanced life support, Resuscitation council- UK 2010. (http://www.resus.org.uk/SiteIndx.htm)

Appendix III
Echocardiographic Reference Values:
M-mode measurements:



BSA (m^2)	0.25	0.3	0.4	0.5	0.6	0.7	0.8	0.9	1.0	1.2	1.4	1.6	1.8	2.0
BW in (kg)[*]	**3**	**4**	**7**	**10**	**13**	**16**	**19**	**23**	**28**	**37**	**46**	**55**	**70**	**80**
LV diastolic dimension	20 (11–29)	22 (13–30)	25 (16–33)	27 (18–37)	30 (21–40)	33 (23–43)	35 (25–45)	37 (27–47)	39 (28–50)	42 (30–53)	43 (31–56)	45 (32–58)	45 (32–61)	46 (32–62)
LV systolic dimension	12 (7–17)	13 (8–19)	15 (9–21)	17 (11–23)	19 (12–25)	20 (14–27)	22 (15–28)	23 (16–30)	24 (17–31)	26 (18–34)	27 (19–36)	28 (10–37)	28 (19–38)	28 (19–39)
IVS thickness (diastolic)	3.5 (1.5–5.5)	3.5 (1.5–5.5)	4 (1.5–6)	4 (2–6.5)	4.5 (2–7)	5 (2–7)	5 (2.5–7.5)	5.5 (2.5–8)	5.5 (3–8.5)	6 (3–9)	7 (3.5–10)	7.5 (4–11)	8 (4–12)	8.5 (4.5–13)
LVPW thickness (diastolic)	3.5 (1.5–5.5)	3.5 (1.5–5.5)	4 (2–6)	4 (2–6.5)	4.5 (2–7)	5 (2.5–7.5)	5 (2.5–7.5)	5 (3–8)	5.5 (3–8.5)	6 (3–9.5)	7 (3.5–10)	7.5 (4–11)	8 (4–12)	8.5 (4.5–13)

Appendix IV

Common Syndromes Associated with Cardiac Involvement

Syndrome	Features	Cardiac Involvement
CHARGE association	*C*oloboma, *h*eart defects, choanal *a*tresia, growth or mental *r*etardation, *g*enitourinary anomalies, *e*ar anomalies, genital hypoplasia	TOF, truncus arteriosus, aortic arch anomalies
DiGeorge syndrome parathyroid, hypocalcemia, deficient cell-mediated immunity	Hypertelorism, short philtrum, downslanting eyes, hypoplasia or absence of thymus and parathyroid, hypocalcemia, deficient cell-mediated immunity	Interrupted aortic arch, truncus arteriosus, VSD, PDA, TOF
Down syndrome (trisomy 21)	Hypotonic, flat facies, slanted palpebral fissure, small eyes, mental deficiency, simian crease	Frequent (40%–50%); AVSD, VSD
Ehlers-Danlos syndrome	Hyperextensive joints, hyperelasticity, fragility and bruisability of skin, poor wound healing with thin scar	Frequent; ASD, aneurysm of aorta and carotids, intracranial aneurysm, MVP
Ellis–van Creveld syndrome (chondroectodermal dysplasia)	Short stature of prenatal onset, short distal extremities, narrow thorax with short ribs, polydactyly, nail hypoplasia, neonatal	Frequent (50%); ASD, single atrium

	teeth	
Goldenhar's syndrome (oculoauriculovertebral spectrum)	Facial asymmetry and hypoplasia, microtia, ear tag, cleft lip/palate, hypoplastic vertebrae	Frequent (35%);VSD, TOF
Glycogen storage disease II (Pompe's disease)	Large tongue and flabby muscles, cardiomegaly; LVH and short PR interval on ECG, severe ventricular hypertrophy on echocardiography	Very common; cardiomyopathy
Holt-Oram syndrome (cardiac-limb syndrome)	Defects or absence of thumb or radius	Frequent; ASD, VSD
Marfan syndrome	Arachnodactyly with hyperextensibility, subluxation of lens	Frequent; aortic aneurysm, aortic and/or mitral regurgitation
Mucopolysaccharidosis	Coarse features, large tongue, depressed nasal bridge, kyphosis, retarded growth, hepatomegaly, corneal opacity (not in Hunter's syndrome), mental retardation; most patients die by 10 to 20 yr.	Frequent; aortic and/or mitral regurgitation, coronary artery disease
Muscular dystrophy (Duchenne's type)	Waddling gait, “pseudohypertrophy” of calf muscle	Frequent; cardiomyopathy
Noonan's syndrome (Turner-like syndrome)	Similar to Turner's syndrome but may occur in phenotypic male and without chromosomal abnormality	Frequent; PS (dystrophic pulmonary valve), LVH (or anterior septal hypertrophy)
Tuberous sclerosis	Triad of adenoma	Frequent;

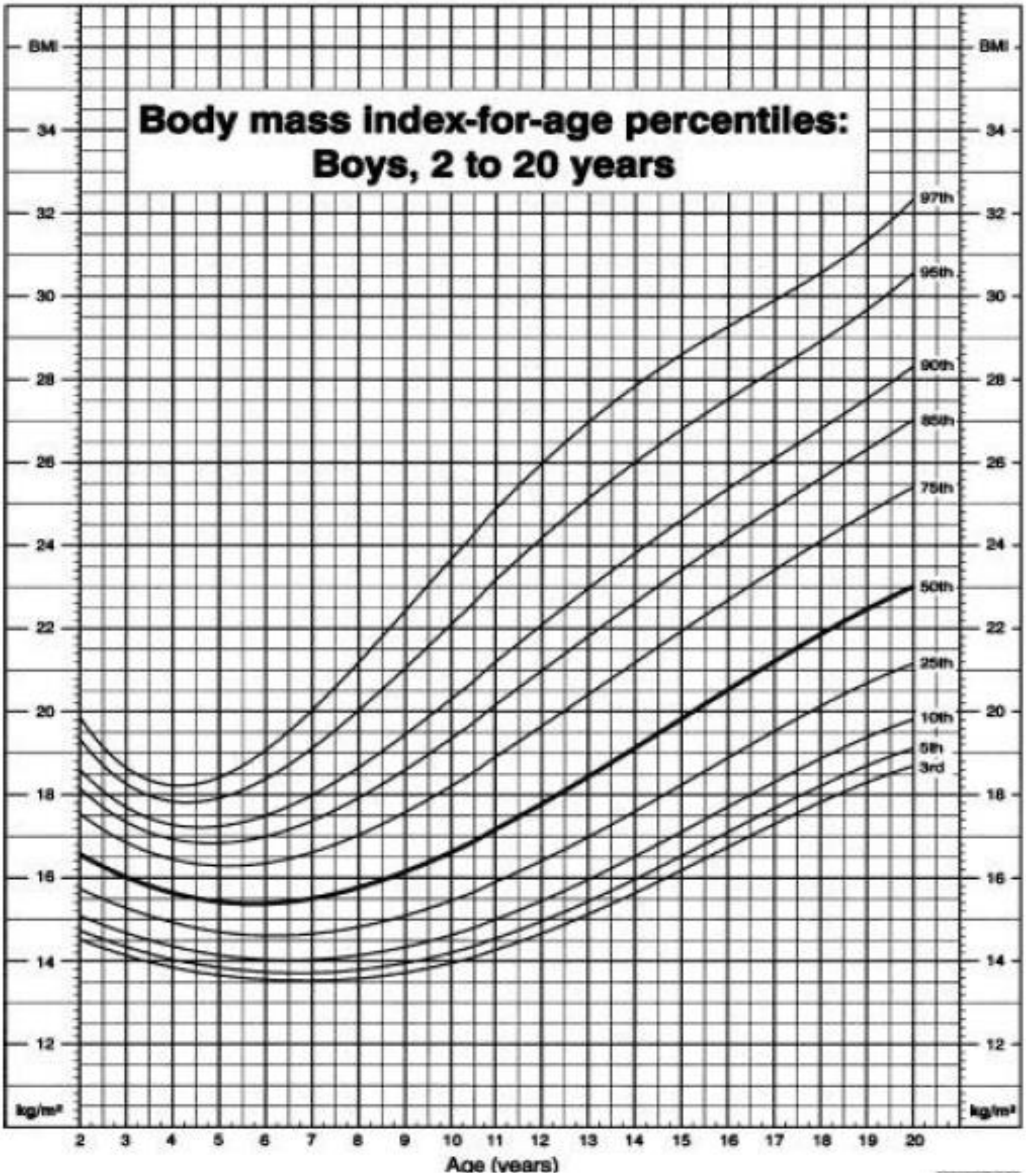
	sebaceum (2–5 yr of age), seizures, and mental defect; cyst-like lesions in phalanges and elsewhere; fibrous angiomatous lesions (83%) with varying colors in nasolabial folds, cheeks, and elsewhere	rhabdomyoma
Turner's syndrome (XO syndrome)	Short female; broad chest with widely spaced nipples; congenital lymphedema with residual puffiness over the dorsa of fingers and toes (80%)	Frequent (35%); COA, bicuspid aortic valve, AS, hypertension, aortic dissection later in life
VATER association (VATER/VACTERL syndrome)	*V*ertebral anomalies, *a*nal atresia, *c*ongenital heart defects, *t*racheo*e*sophageal (TE) fistula, *r*enal dysplasia, *l*imb anomalies (e.g., radial dysplasia)	Common (>50%); VSD, other defects
Williams syndrome	Varying degrees of mental retardation, so-called elfin facies (consisting of some of the following: upturned nose, flat nasal bridge, long philtrum, flat malar area, wide mouth, full lips, widely spaced teeth, periorbital fullness), hypercalcemia of	Frequent; supravalvular AS, PA stenosis

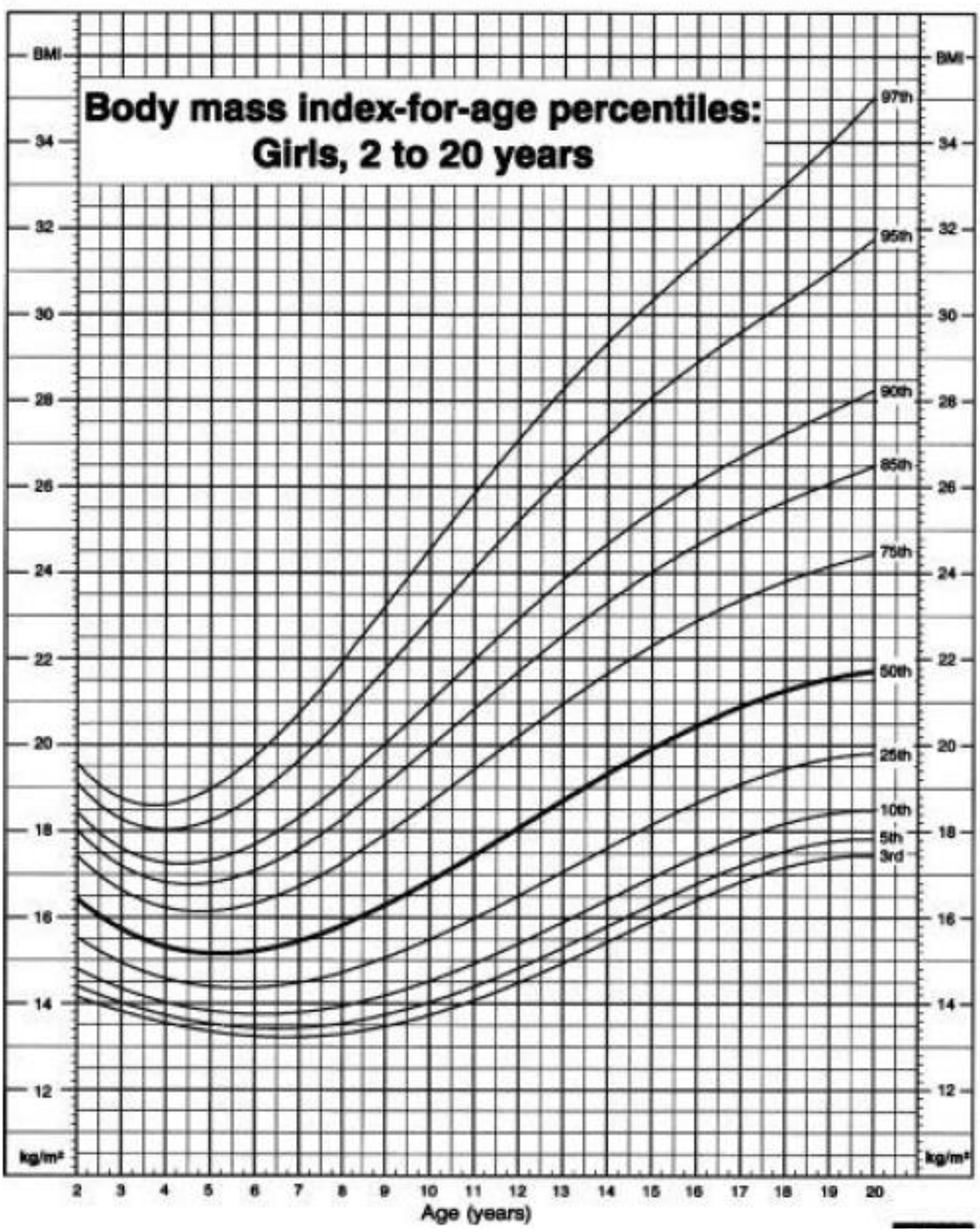
	infancy?	
Zellweger's syndrome (cerebrohepatorenal syndrome)	Hypotonia, high forehead with flat facies, hepatomegaly, albuminuria	Frequent; PDA, VSD or ASD

Modified from Park MK. Basic Tools in Routine Evaluation of Cardiac Patients. In: Pediatric cardiology for practitioners.5th ed. Philadelphia, Mosby, Elsevier2008.

Appendix V

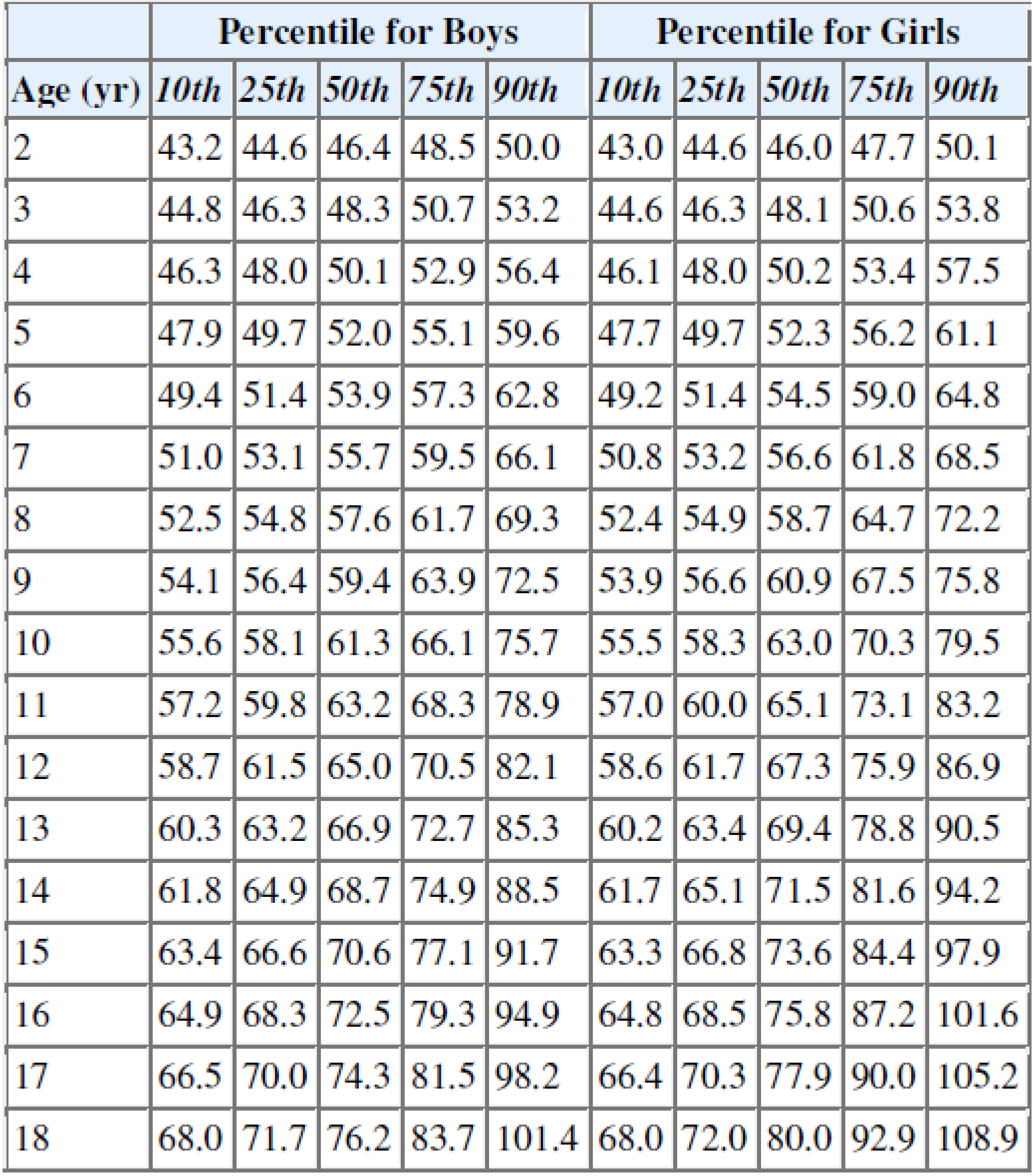
Body Mass Index for Children (Boys and Girls)




Body mass index-for-age percentiles:
Girls, 2 to 20 years
BMI
34
32
30
28
26
24
22
20
18
16
14
12
kg/m²
97th
95th
90th
85th
75th
50th
25th
10th
5th
3rd
2
3
4
5
6
7
8
9
10
11
12
13
14
15
16
17
18
19
20
Age (years)

Appendix VI

Waist Circumference for Children and Adolescents according to sex:



	Percentile for Boys					Percentile for Girls				
Age (yr)	*10th*	*25th*	*50th*	*75th*	*90th*	*10th*	*25th*	*50th*	*75th*	*90th*
2	43.2	44.6	46.4	48.5	50.0	43.0	44.6	46.0	47.7	50.1
3	44.8	46.3	48.3	50.7	53.2	44.6	46.3	48.1	50.6	53.8
4	46.3	48.0	50.1	52.9	56.4	46.1	48.0	50.2	53.4	57.5
5	47.9	49.7	52.0	55.1	59.6	47.7	49.7	52.3	56.2	61.1
6	49.4	51.4	53.9	57.3	62.8	49.2	51.4	54.5	59.0	64.8
7	51.0	53.1	55.7	59.5	66.1	50.8	53.2	56.6	61.8	68.5
8	52.5	54.8	57.6	61.7	69.3	52.4	54.9	58.7	64.7	72.2
9	54.1	56.4	59.4	63.9	72.5	53.9	56.6	60.9	67.5	75.8
10	55.6	58.1	61.3	66.1	75.7	55.5	58.3	63.0	70.3	79.5
11	57.2	59.8	63.2	68.3	78.9	57.0	60.0	65.1	73.1	83.2
12	58.7	61.5	65.0	70.5	82.1	58.6	61.7	67.3	75.9	86.9
13	60.3	63.2	66.9	72.7	85.3	60.2	63.4	69.4	78.8	90.5
14	61.8	64.9	68.7	74.9	88.5	61.7	65.1	71.5	81.6	94.2
15	63.4	66.6	70.6	77.1	91.7	63.3	66.8	73.6	84.4	97.9
16	64.9	68.3	72.5	79.3	94.9	64.8	68.5	75.8	87.2	101.6
17	66.5	70.0	74.3	81.5	98.2	66.4	70.3	77.9	90.0	105.2
18	68.0	71.7	76.2	83.7	101.4	68.0	72.0	80.0	92.9	108.9

From Fernandez JR, Redden DT, Pietrobelli A, Allison DB: Waist circumference percentiles in nationally representative samples of African-American, European-American, and Mexican-American children and adolescents. J Pediatr 145:439–444, 2004.